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ABSTRACT

Presented in the conference proceedings are schedule and list of participants, seven major papers, and the newborn hearing screening recommendations of the interdisciplinary conference on newborn hearing and early identification of hearing impairment. Neonatal auditory testing is reviewed by Sanford E. Gerber, and Sheldon B. Korones gives a neonatologist's overview of screening programs for detecting deafness in newborns. Neurosensory factors in newborn hearing are considered by Louis Gluck, while Bruce W. Konigsmark discusses hereditary and congenital factors affecting newborn sensorineural hearing and Burton F. Jaffe discusses hereditary and congenital factors affecting conductive hearing. Arthur J. Derbyshire presents theoretical considerations in the selection of variables for testing newborn hearing, and, in conclusion, a current overview of newborn hearing screening is given by Marion P. Downs. (KW)

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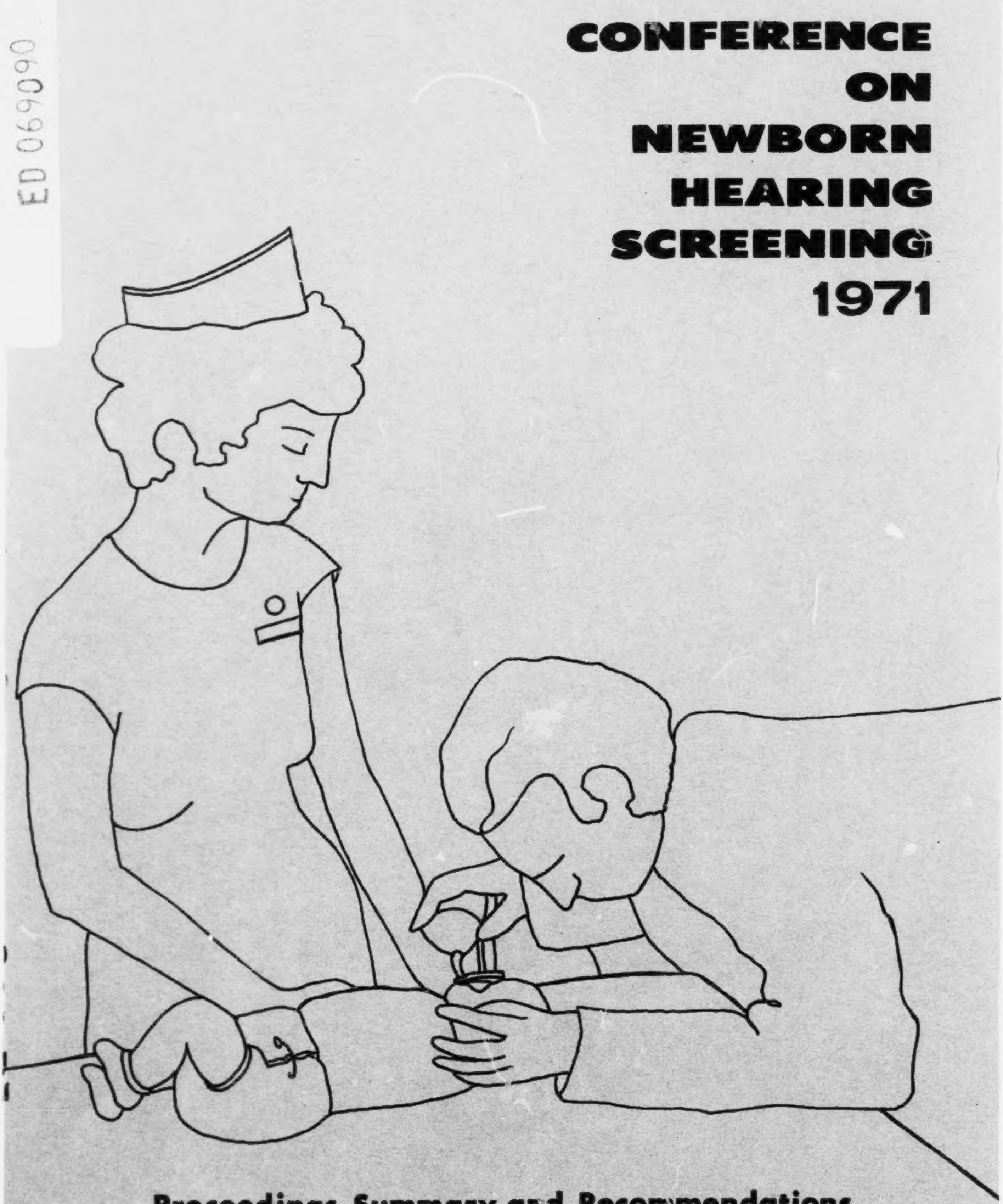
# CONFERENCE ON NEWBORN HEARING SCREENING 1971



## Proceedings Summary and Recommendations

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# CONFERENCE ON NEWBORN HEARING SCREENING

## Proceedings Summary and Recommendations

SAN FRANCISCO INTERNATIONAL AIRPORT
HILTON INN
CALIFORNIA

FEBRUARY 23, 24, 25, 1971

CONFERENCE SPONSORS

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# INTRODUCTION

The publication of these proceedings of a conference on Newborn Hearing and Early Identification of Hearing Impairment held on February 23 - 25, 1971 at the San Francisco Airport represents the culmination of more than a year of effort of a number of interested agencies and individuals, from clinical, academic and public health orientations.

The germination of the idea of an interdisciplinary top-level conference began during a tri-regional meeting sponsored by the Health Services and Mental Health Administration of Health, Education, and Welfare, relating to the problem of earlier detection and treatment of handicapping conditions in children in early 1970. During the discussion there it became evident that there existed areas of controversy about hearing screening of the newborn. We in the California State Department of Public Health, Bureau of Maternal and Child Health offered to co-sponsor such a meeting with the Maternal and Child Health Services of the Health Services and Mental Health Administration, Department of Health, Education, and Welfare, and with assurance of the advice and counsel of the Joint Committee on Newborn Screening representing the American Academy of Pediatrics, the American Academy of Ophthalmology and Otolaryngology, and the American Speech and Hearing Association.

The Bureau of Maternal and Child Health conducted a mail-response opinion poll on the practices and opinions of audiologists, pediatricians and otologists relating to infant auditory screening.* The results of the poll were encouraging, and planning for the conference proceeded, with the added assurance from invited speakers and participants of their interest and willingness to contribute.

Objectives set for the conference were:

1. To compile, assess, summarize, and report information related to neonatal hearing and its relationship to other aspects of growth and development.
2. To assess the desirability, practicability, and significance of each of a variety of hearing testing procedures for the newborn's hearing.
3. To develop guidelines related to testing the newborn's hearing.
4. To recommend studies which should be conducted related to the newborn's responses to acoustic stimuli with a view to develop increasingly effective means for early identification of hearing impairment.

The conference was designed to maximize both formal and informal opportunity for information and opinion exchange among the participants. The number of invited participants was limited to keep the working sessions of manageable size. The first day's sessions, devoted as they were primarily to the presentation of the papers, were opened to the professional public. The second day was given over to small group discussions and reporting back to the entire conference. The conferees met once more in a morning session to review and agree on the recommendations arising from the previous day's work.

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* Questionnaire and results of poll are included in Appendices I and II, pages 133-134

The contents of the published proceedings include all formal papers, as prepared originally or as edited by the authors following the meeting in the interests of clarity, and the summarized recommendations.

Special acknowledgments are due the Joint Committee on Newborn Screening under the chairmanship of Mrs. Marion P. Downs for their initial interest in getting the conference going and their considerable individual contributions of time and talent as speakers, chairmen and prime movers. To John J. Hutchings, M.D. and Don Harrington, Ph.D. of Maternal and Child Health Services, Health Services and Mental Health Administration, go special thanks for their smoothing of fiscal and administrative paths in funding and organizing the conference.

George Cunningham

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May 14, 1971

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# CONFERENCE ON NEWBORN HEARING AND EARLY IDENTIFICATION OF HEARING IMPAIRMENT

## AGENDA

Revised

February 23*

8:00 - 8:30 Registration

8:30 - 9:00 Welcome

Hamlet C. Pulley, M.D.
Assistant Director
California State Department of Public Health

John J. Hutchings, M.D.
Regional Medical Director
Maternal and Child Health Service
Department of Health, Education, and Welfare

"The MCH Perspective"

George C. Cunningham, M.D.
Chief, Bureau of Maternal and Child Health

9:00 - 4:00 Chairman: Sheldon Korones, M.D.

9:00 - 9:10 "The Pediatric Overview of Newborn Hearing"
Paper: Screening Programs for the Detection of Deafness in Newborn Infants – A Neonatologist's Overview

Sheldon Korones, M.D.

9:10 - 10:00 Neurosensory Factors in Newborn Hearing

Louis Gluck, M.D.

10:00 - 10:15 BREAK

10:15 - 11:15 Hereditary and Congenital Factors Affecting Newborn Sensorineural Hearing

Bruce Konigsmark, M.D.

11:15 - 12:15 Hereditary and Congenital Factors Affecting Newborn Conductive Hearing

Burton F. Jaffe, M.D.

12:15 - 1:15 LUNCH

1:15 - 2:15 Theoretical Considerations in the Selection of Variables for Testing the Hearing of Newborns

A. J. Derbyshire, Ph.D.

2:15 - 2:30 BREAK

2:30 - 4:00 Discussion of all four papers

February 24, 1971

8:30 - 9:00 General Workshop Session: Vintage Rooms 7 and 8

Chairman: Mrs. Downs

Current Overview of Newborn Hearing Screening –

Mrs. Marion P. Downs

9:00 - 12:00 Group Discussions

Group I - Directors' Room 600

Dr. G. Cunningham, Chairman
Dr. W. Hawes, Recorder

Group II - Directors' Room 602

Dr. D. Harrington, Chairman
Mr. D. Caziarc, Recorder

Group III - Directors' Room 604

Mrs. M. Downs, Chairman
Miss G. Church, Recorder

12:00 - 1:00 LUNCH

1:00 - 2:45 Group Discussions continue

2:45 - 3:00 BREAK

3:00 - 4:00 General Workshop Session: Vintage Rooms 7 and 8

Chairman: Dr. Harrington

Presentation of each group's recommendations by the group chairmen and recorders.

February 25, 1971

8:30 - 12:00 General Workshop Session: Vintage Rooms 7 and 8

Chairman: Dr. Cunningham

Formulation of statements upon which there is substantial agreement which can become the guidelines for Federal and State agencies.

Entire group will consider the group recommendations. Similar items from each report will have been grouped together prior to this meeting to facilitate discussion.

## NEWBORN HEARING SCREENING RECOMMENDATIONS

I would like to preface these recommendations by saying that we were most fortunate to have had the opportunity to assemble such a qualified and experienced group of professionals from a broad spectrum of disciplines. I believe that we have gone far toward accomplishing the conference objectives and the proceedings of this conference will be a most valuable resource for others who will consider this same subject in the future.

The formulation of general statements and recommendations was perhaps the most difficult task facing the Conference. In selecting the participants we made a deliberate effort to include both proponents and opponents of mass newborn screening and realized that as a result the differences of opinion and the broad spectrum of points of view represented would not be conducive to unanimity of conclusions.

But the situation is not at all bleak; while we found the diversity we expected, we also found some broad areas of agreement. The statements listed under "Recommendations" were then submitted to the assembled group and approved after general discussion. Participants were given an opportunity to review the written recommendations subsequent to the meeting. The following statements represent the final recommendations of the conference.

George C. Cunningham

## RECOMMENDATIONS

I. **A HIGH RISK POPULATION CAN AND SHOULD BE IDENTIFIED BY PRENATAL HISTORY AND POSTNATAL PHYSICAL ASSESSMENT OF THE INFANT. AS A FIRST STEP A REGISTRY SHOULD CONTAIN THE FOLLOWING GROUPS.**

**PRENATAL HIGH RISK PROCEDURE**

1. **ALL INFANTS WITH A FAMILY HISTORY OF CHILDHOOD DEAFNESS IN SOME MEMBER OF THE IMMEDIATE FAMILY, I.E., FATHER, MOTHER, OR SIBLING.**
2. **ALL INFANTS WHOSE MOTHERS HAVE HAD RUBELLA DOCUMENTED OR STRONGLY SUSPECTED DURING ANY PERIOD OF PREGNANCY.**
3. **ALL INFANTS WITH A FAMILY HISTORY OF CONGENITAL MALFORMATIONS OF THE EXTERNAL EAR, CLEFT LIP OR PALATE.**
4. **ALL INFANTS WITH A FAMILY HISTORY OF DEAFNESS IN OTHER RELATIVES, WITH ONSET IN CHILDHOOD.**

POSTNATAL

5. ALL INFANTS FOUND TO HAVE A STRUCTURAL ABNORMALITY OF THE EXTERNAL EAR, CLEFT LIP OR PALATE, INCLUDING BIFID UVULA.

6. ALL INFANTS HAVING BILIRUBIN VALUES OF 20 MG/100 MG OR MORE, WHO HAD EXCHANGE TRANSFUSIONS ARE AT HIGH RISK OF BILIRUBIN ENCEPHALOPATHY.

7. ALL INFANTS UNDER 1,500 GRAMS.

8. ALL INFANTS WITH ABNORMAL OTOSCOPIC FINDINGS.

*(It was suggested that Groups 1, 2 and 5 would be referred to an otologic-audiologic testing center for more elaborate workup by available methods, such as evoked potential, cardiac response audiometry and followed by periodic followup evaluations including consecutive tests.)*

II. WHILE FROM THE STANDPOINT OF PREVENTIVE, THERAPEUTIC AND REHABILITATIVE INTERVENTION THERE IS LITTLE SPECIAL ADVANTAGE TO DETECTING CONGENITAL DEAFNESS IN THE FIRST FEW DAYS OF LIFE, AND ACKNOWLEDGING THAT SCREENING TEST VALIDITY AND ACCURACY ARE IMPROVED IF SCREENING IS DEFERRED TO A LATER AGE, THE POSSIBILITY OF SCREENING ALL NEWBORNS SHOULD BE SERIOUSLY CONSIDERED IF: (1) ONE OR MORE RELATIVELY RELIABLE AND INEXPENSIVE AUDITORY SCREENING TECHNIQUES ARE AVAILABLE, AND (2) IT CAN BE SHOWN THIS PROCEDURE YIELDS A SIGNIFICANT NUMBER OF CASES THAT WOULD BE MISSED OR NOT DETECTED UNTIL AFTER THE OPTIMAL PERIOD FOR INTERVENTION IF AN ALTERNATE PROGRAM WERE SELECTED FOR SCREENING BASED ON DELAYED TESTING ALONE.

III. AN AUDITORY SCREENING TECHNIQUE, IF IT IS TO BE APPLICABLE TO ALL INFANTS, MUST BE SIMPLIFIED SO AS TO MEET THE FOLLOWING CRITERIA AS FAR AS POSSIBLE. IT SHOULD

1. REQUIRE A MINIMAL INVESTMENT IN NEW PERSONNEL, TRAINING, TIME AND EQUIPMENT;

2. DETECT A SIGNIFICANT NUMBER OF INFANTS WITH IMPAIRED HEARING THAT WOULD NOT BE DETECTED BY HIGH RISK SCREENING ALONE;

3. HAVE AN ACCEPTABLY LOW RATE OF FALSE NEGATIVES (MISSED CASES) AND FALSE POSITIVES;

4. HAVE A SCREENING CRITERIA THAT IS CLEARLY PASS OR FAIL WHETHER BY SUBJECTIVE JUDGMENT, E.G., AWAKENS VERSUS DOES NOT AWAKEN, OR BY INSTRUMENT RESULTS, E.G., RED LIGHT, GREEN LIGHT, AMBER LIGHT.

IV. BECAUSE OF LIMITED RESOURCES AVAILABLE TO THE HEALTH DELIVERY SYSTEM, AN ANALYSIS OF THE NET COST VERSUS NET BENEFIT OF ANY PROPOSED NEWBORN SCREENING PROGRAM AND THE RELATIVE PRIORITY FOR THIS INVESTMENT OF RESOURCES MUST BE CONSIDERED. DATA SHOULD BE ASSEMBLED AND DEVELOPED THAT COULD BE USED TO ILLUSTRATE THE NET ADVANTAGE TO THE INDIVIDUAL AND SOCIETY OF EARLY CASE FINDING BEFORE EARLY DETECTION CAN BE RECOMMENDED AS A HIGH PRIORITY ITEM FOR HEALTH FUNDS. IN CONSIDERATION OF THESE COST FACTORS THE SCREENING OF ONLY HIGH RISK REGISTRY INFANTS SHOULD BE ANALYZED AND MIGHT BE ACCEPTABLE AS AN ALTERNATIVE TO UNIVERSAL SCREENING.

V. REGARDLESS OF WHAT IS DONE IN THE NURSERY, FOLLOWUP INCLUDING REEXAMINATION BY A SUITABLY RELIABLE TECHNIQUE MUST BE PART OF THE RECOMMENDED PROGRAM FOR ALL INFANTS DURING THE FIRST YEAR OF LIFE.

VI. PARENTS AND ALL HEALTH AND EDUCATIONAL PERSONNEL SHOULD BE ENCOURAGED TO REFER ALL SUSPECTED CHILDREN AS THEY CAN BE TESTED AT ANY AGE. THERE IS NO AGE TOO YOUNG FOR DETECTION OR FOR REHABILITATION. IN IMPLEMENTING THIS RECOMMENDATION AN ASSESSMENT OF RESOURCES AVAILABLE FOR SERVICES TO THOSE DETECTED AND REFERRED SHOULD FIRST BE MADE.

VII. THE CONFERENCE RECOMMENDS THAT GREATER EFFORT BE MADE TO TRAIN THOSE RESPONSIBLE FOR THE CARE OF INFANTS IN THE EVALUATION OF THE EAR WITH THE PNEUMATIC OTOSCOPIC AND ENCOURAGES SUCH EVALUATIONS OF INFANTS AND TODDLERS. SUCH EVALUATION SHOULD INCLUDE A BRIEF HISTORY AND AN APPROPRIATE TEST OF AUDITORY FUNCTION PERFORMED WITH SIMPLE EQUIPMENT BY SUITABLY TRAINED PERSONNEL. SCREENING PROGRAMS SHOULD BE DESIGNED TO UTILIZE THE SKILLS AND RESOURCES OF PUBLIC HEALTH AGENCIES IN ASSURING COMPLETENESS OF FOLLOWUP.

VIII. ALL SCREENING PROCEDURES SHOULD BE VIEWED AS PART OF COMPREHENSIVE HEALTH CARE AND ULTIMATELY COORDINATED INTO AN EFFICIENT MULTIFACETED SCREENING AND HEALTH SURVEILLANCE AND MAINTENANCE PROGRAM.

IX. AREAS IDENTIFIED AS PROMISING POSSIBILITIES FOR RESEARCH EFFORTS THAT COULD CONTRIBUTE TO THE DECISION TO SCREEN AND TO THE DESIGN OF THE SCREENING PROGRAM ARE:

1. VALIDATION OF THE CRIBOGRAM APPROACH ON A LARGE SAMPLE IN SEVERAL INSTITUTIONS WITH APPROPRIATE FOLLOWUP;
2. VALIDATION OF A SIMPLE AWAKENING RESPONSE TO A SPECIFIED STANDARD STIMULUS (90 dB fluctuating 2,000-3,000 Hz. noise at 14 inches) WITH APPROPRIATE FOLLOWUP;
3. SIMPLIFICATION OF THE INSTRUMENTAL (OBJECTIVE) APPROACH TO SCREENING;
4. LONG-TERM PERIODIC TESTING OF A HIGH RISK POPULATION PROSPECTIVELY TO ASCERTAIN THE FREQUENCY OF ACQUIRED DEAFNESS;
5. EFFORTS SHOULD BE MADE TO IDENTIFY CONDITIONS WHICH ARE ASSOCIATED WITH A HIGH RISK OF HEARING PROBLEMS TO BE ADDED TO THE HIGH RISK REGISTRY; CRITERIA OF HIGH RISK SHOULD BE REFINED BY A CONTINUING INVESTIGATION OF THE CORRELATION BETWEEN SELECTED PHENOMENA AND HEARING DEFECTS;
6. EVALUATION SHOULD BE MADE OF THE USEFULNESS AND RELIABILITY OF VESTIBULAR TESTING IN THE NURSERY AND DURING INFANCY;
7. THERE SHOULD BE EPIDEMIOLOGICAL STUDIES OF THE INCIDENCE AND CLINICAL SIGNIFICANCE OF CONDUCTIVE LOSSES IN THE FIRST YEAR OF LIFE;
8. THE EFFECTIVENESS OF GENETIC COUNSELING IN VARIOUS SETTINGS ON FAMILY SIZE SHOULD BE EVALUATED;
9. EVALUATION OF THE AVAILABLE HEARING EVALUATION TESTS FOR EARLY INFANCY (4 TO 8 MONTHS) SHOULD BE CARRIED OUT AND A SIMPLE PRACTICAL PROCEDURE RECOMMENDED AND PROMOTED.

# NEONATAL AUDITORY TESTING: A REVIEW

Sanford E. Gerber, Ph.D.*

It has been a most illuminating experience to collect the literature on the subject of neonatal auditory testing for this conference. Following this review is an extensive bibliography which has caused the writer to become ever more aware of the subject matter. In fact, it was amazing what a large body of literature exists on this subject. Even though there is such a large body of literature, it is even more amazing how ignorant we remain. It is this ignorance, along with our abiding concern, that has caused our Joint Committee on Newborn Hearing Screening Programs to generate the following statement:

> In recognition of the need to identify hearing impairment as early in life as possible, auditory screening programs have been implemented in newborn nurseries throughout the country. Review of data from the limited number of controlled studies which have been reported to date has convinced us that results of mass screening programs are inconsistent and misleading.
>
> To determine whether mass screening programs for newborn infants should indeed be instituted, intensive study of a number of variables is essential. These should include stimuli, response patterns, environmental factors, status at the time of testing, and behavior of observers. Furthermore, confirmation of results obtained in the nursery must await data derived from extended follow-up studies which involve quantitative assessment of hearing status.
>
> In view of the above considerations and despite our recognition of the urgent need for early detection of hearing impairment, we urge increased research efforts, but cannot recommend routine screening of newborn infants for hearing impairment.

In preparing this review of the literature, I came much more fully to an understanding of the preceding statements which I have endorsed. It was this statement which I used to provide an outline for the review which follows. Following an introductory section and a statement of need, the outline proceeds as suggested by the committee statement. It will consider maturation, stimuli, response patterns, environmental factors, infant status, behavior of observers and what little data we could find on follow-up studies. After this review will be found the lengthy bibliography mentioned above.

---

* Associate Professor of Audiology, Speech and Hearing Center, University of California, Santa Barbara.

## INTRODUCTION AND BACKGROUND

In this first section of our review we want to consider those things which have led us to think we might screen the hearing of neonates at all. Reasons why we should are in the next section. One finds in the literature a number of statistical studies showing how many newborn infants have been tested for hearing or hearing loss and how many hearing impaired infants have been found in this manner. Downs (1965) in reviewing her experiences said: "Experience in observing the auditory behavior of over 5,000 newborn infants leads us to believe that it is feasible to screen for peripheral hearing deficits at birth . . ." Essentially the same observation was made by Redell (1970): "Our experience in screening more than 5,000 newborns demonstrates that their hearing can be tested simply, quickly, and effectively in the hospital nursery." Redell went on to recommend that such screening ought to be led by the thinking of pediatricians. In 1969 Downs and Hemenway reported on the screening of 17,000 newborn infants. Their statistical report indicated the incidence of hearing loss at birth to be one in 1,000. A paper by Wedenberg in 1963 indicated that such neonatal testing does render reliable results. One might conclude from the foregoing that such a concept is rather recent. That is not the case, however. As early as 1928 Aldrick recommended a test for hearing in newborns. He observed that he could pair the ringing of a bell with scratching the bottom of a foot of a three month old infant. This elicited a conditioned reflex (in the Pavlovian sense) as the infant would pull the foot away when this bell was heard. Aldrick proposed that a test of this kind could be used to test the hearing of newborns. Similarly, Froeschels and Beebe (1946) reported significant findings on newborn infants. The general observation from studies of this type is that it *is* possible to elicit auditory responses from neonates. In fact, the Committee on Child Health of the American Public Health Association (1956) suggested that hearing testing may be done with infants as young as ten days, and Downs in 1967 (a) prepared a manual describing how a newborn screening program could be set up, who should do the testing, where testing ought to be done and how, and how to interpret results, as well as how to train observers and do follow-up studies.

Nevertheless, some other brilliant and talented observers have had an amount of success of less satisfaction than that of others. Janet Hardy (1965), for example, concluded: "In my opinion, testing of the newborn as we have been doing it is useless and we plan to discontinue it . . ." Certainly there are difficulties in testing neonates which may be obviated if the tester waits for the infant to get a little older. Fisch (1970) observed: "It is not certain that a subject, even with normal hearing, will give a reaction to a sound stimulus every time . . ." Similarly, Trenque (1962) concluded that "No useful purpose could be served" by an attempt to determine anything other than what he called "audio-estimation" for the newborn infant. Part of this difficulty, of course, has been in determining what is our goal. The present author asked this question in somewhat different terms (Gerber, 1969). At that time I said: "It may be easy to recognize the infant who does not respond, but what about the one who does? What does a hearing baby do to indicate that fact? Or, to phrase it differently, what does the responsive infant do that the unresponsive infant does not do?" Perhaps the most significant observation was that of William Hardy (1965). He told us: "It is important in the assessment of tiny infants to understand exactly what is being attempted. The tester is not trying to diagnose deafness. He is presenting certain stimuli and observing the responses. Either the baby responds in a way that the observers can see and agree on, or he does not. Some responses are repeatable, others are not. If a baby does not respond, one cannot conclude that he does not *hear*; only that he does not *respond*."

One may see, therefore, that there is not a consensus on the possibility of testing the hearing of newborn infants. Most experienced persons agree, however, that such can be done under certain circumstances but their opinions need to be tempered by the considered observations of some major contributors to this area. One does arrive at the rather early conclusion of Hardy and Bordley (1951): "By and large, however, all such testing fails to measure auditory acuity or sensitivity with sufficient refinement; it is rather much an open-or-shut matter without quantification." Earlier (Gerber, 1969) I made essentially the same observation by pointing out that the kind of screening we do perhaps may discriminate the deaf child from the child who is not deaf, but cannot discriminate the deaf child from the child who is hard of hearing, or the hard of hearing child from the child with normal hearing.

## STATEMENT OF NEED

This review would not exist if there were not some kind of consensus that it is possible to test the hearing of the newborn. Of even greater significance is the consensus that it is important and even necessary to test the hearing of the newborn. In the previous section we pointed out that Downs and Hemenway (1969) found the incidence of hearing loss at birth to be one in 1,000 in their study of 17,000 newborn infants. In another paper by Mrs. Downs (1968) she suggested an incidence figure approaching one in 1,500 infants. A paper published anonymously in Canada (1967) pointed out that after a screening of 6,000 newborn infants, three were shown to have hearing loss. That means, then, an incidence of one in 2,000. One can conclude from these statistical studies very generally that the incidence of congenital deafness ranges from one in 1,000 to one in 2,000, perhaps depending upon how deafness is defined and how it is determined. A very conservative estimate (Downs and Sterritt, 1967) found that peripheral hearing impairment may appear in one of every 2,000 or 4,000 babies. The problem of confusing peripheral with central auditory disorders was discussed by Kimball in 1967. He observed that infants with central disorders may give reflexive responses to acoustic stimuli and be passed as normal; or, if they do not respond to acoustic stimuli, they may be misdiagnosed as peripherally deaf.

In addition to the statistical factors raised, there are other reasons discussed by some authors as to why it is necessary to screen at birth. Duffy (1962) pointed out that early detection is important to determine prognosis. Fisch (1964) considered the necessity of normal hearing in the first six months of life for an infant to relate satisfactorily to his environment. Flower (1968) also pointed out ". . . these observations should begin in the hospital nursery." Similarly, Kendall (1965) said: "No child is too young to be tested, and screening techniques can be used successfully with newborns." Kendall was concerned that diagnosis and treatment "depend upon vigorous, early and effective case finding." He was fearful that not testing at birth may lead to needless delay and "such delay may compromise the whole development of the hearing impaired child." Essentially the same observation was made by Matkin (1968) who observed ". . . it is apparent that the pervasive effects of an auditory deficit can be successfully minimized only if an aggressive habilitation program is initiated at an early age." He therefore concluded that such auditory screening ought to be undertaken "either in the hospital nursery or during the routine office examination . . ." A most serious observation was published in 1965 by Røjskjaer who noted ". . . some recent biological experiments indicate that complete sensory deprivation during the first months of life may interfere with the normal development and organization of sensory systems. If this theory is correct, then an early diagnosis may be very important." Certainly the general conclusion to be derived from investigation of the need is that hearing losses and other neurosensory deficits do exist at birth and frequently can be identified in the newborn nursery. There has been some question, however, if it is necessary to introduce screening programs for this purpose. Downs (1968) considered the possibility of screening only high risk babies. Robinson (1965) has also pointed out that hearing losses have causes and it is possible to be aware of these causes. A 1966 paper of Eisenberg, Coursin and Rupp concluded that differential responses may be observed if newborns are categorized on the basis of risk. It has similarly been observed that premature, well born, and normal neonates respond differentially to acoustic stimuli (Field, et al., 1967). Many high risk registers have been developed for the purpose of predicting those infants who might have auditory or other neurosensory deficits. It is anticipated by some that the high risk register, if it is successful, obviates the need for mass screening. Certainly errors can be made in mass screening. Downs (1967b) discovered: "A fairly high rate of false positives is found in the program: one and one-half per cent of the total population failed to respond to the test signal, and 97 per cent of these are ultimately cleared." Similarly, Ling, Ling, and Doehring (1970) in investigating their own screening program concluded that ". . . many of the observed responses to sound appear to be of doubtful validity."

Nevertheless, there seems to be general consensus that we do need some kind of auditory screening or auditory high risk register at the time of birth. One of the principal reasons for this consensus, and one which underlay the recommendations of the Joint Committee on Neonatal Hearing Screening Programs, is that in many parts of the country children are examined by physicians only at birth. Many of us are concerned that failure to examine the child at birth may imply failure to examine the child ever. In that case it is probable that a significant proportion of genuinely hearing impaired children will never be diagnosed as such. This observation was made, in a sense, by Fisch in his 1964 monograph to the effect that newborns are available, thus permitting nearly 100 per cent case findings. A similar observation was made by Barr in 1965 to the effect that today 99 per cent of the children born in Sweden are born in hospitals. It is in those hospitals where the screening can be done. It has also been observed by Downs and Sterritt (1964) that very high interobserver reliability could be obtained. Such reliability, however, need not imply validity. In a paper by Eisenberg (1965b) she pointed out that high interobserver reliability means only that the observers are responding to the same behavior. It does not imply necessarily that the behavior which they observe is an auditory response. Ling, Ling, and Doehring (1970) suggested that: "Observers are likely to accept behavioral changes arising for other reasons as positive responses." These observations cause us to repeat Hardy's (1965) comment that "If a baby does not respond, one cannot conclude that he does not *hear*; only that he does not *respond*."

Several reasons have been educed as to why one ought to screen the hearing of neonates. These reasons include the fact that it may be the only possible time to do so, that experiences of several persons have been that auditory and other neurosensory deficits are present and observable at birth, and that auditory deficits identifiable at birth require the earliest possible habilitation. On the other hand, some authorities have suggested that a number of errors, both positive and negative, which can be made may outweigh the apparent benefits of such a program.

## MATURATION

All of the preceding implies that the human neonate has achieved a level of maturation so that we may expect him to respond to acoustic stimulation. This has not always been the case. It is of especial interest that our colleagues, the otolaryngologists, seem to have been the last to recognize this. In 1932, Phillips and Rowell said: "We are all born deaf as well as dumb." However, the preceding year Marquis (1931) had demonstrated that newborn infants can be conditioned to the sound of a buzzer. But perhaps those particular otolaryngologists were just a bit behind the times. In 1907, Barnhill and Wales in their text on otology said: "In the infant hearing is the last sense to awaken." They were confident that no reaction to sound could be elicited at least during the first three days of life. But in 1882 Genzmer claimed that neonates do hear on the first or certainly the second day of life and probably also in utero. Perhaps the earliest raising of this question was in 1859 by Kussmaul who found no evidence of auditory sensitivity in the newborn. It does appear, however, that somewhere between 1859 and 1882 experimenters were interested in this kind of behavior and discovered that indeed it does exist. During the early 30's (Phillips and Rowell notwithstanding) many investigators were not only convinced that neonates respond to sound but were interested to what extent they had matured with reference to the sounds to which they might make responses. The nature of neonatal auditory responses and stimuli which elicit them was investigated by Pratt, Nelson and Sun in 1930, by Marquis in 1931, by Haller in 1932 and by Stubbs in 1934. So we see that during the early 30's the subject of auditory behavior in the neonate and the matter of neonatal auditory maturation had certainly attracted substantial attention.

Today there is evidence that the human auditory nervous system is functioning not only at birth but indeed before birth. This paper does not deal specifically with prenatal auditory behavior but the reader may be interested in seeing the study of Johansson, Wedenberg, and Westin (1964). Studies of premature infants showing the nature of their auditory behavior at birth necessarily lead one to the conclusion that the auditory sensory system is intact and operating prior to birth in a well born infant. This is shown by data such as those gathered by Vasiliu (1968): "Faced with these findings we ascertained that the child is ready to hear, even in the intrauterine stage. This is proved by the fact that the premature child of six to seven months can hear after birth." Similar observations were made by that master pediatrician, Arnold Gesell. In 1947, in the second edition of their book, Gesell and Amatruda said: "There is good evidence that the fetus responds with sudden movements to loud sounds . . . certain it is that premature infants with a fetal age of 30 weeks or more . . . will react positively to the sound of a tinkled bell . . ." The study of the "neurological maturation of the child, whatever age he may be, from the barely viable premature baby to the stage of autonomous infant . . ." permits one to generate criteria of maturation (Dargassies, 1969). Development of such maturational criteria has been studied in premature babies of conceptional age 24 to 41 weeks (Dreyfus-Brissac, 1969). Again, the auditory behavior can be observed at this point.

Indeed the healthy full term infant displays a number of responses to auditory stimuli very very early in his life.* Yang (1962) observed the Moro reflex on the first day of life. Wertheimer in 1961 showed that infants only a few minutes old turn their eyes in the direction of auditory stimuli. It is also possible to observe the maturation of auditory behavior still within the neonatal period. Gesell and Amatruda (1947) pointed out "the full term neonate assumes nothing less than listening attitudes to the sound of the human voice, within the first fortnight after birth." In 1969, Murphy showed that what is apparently the auditory attention of the newborn modifies during the first week of life. He suggested that at the end of that first week "A generalized diminution of sensory function is noted . . ." He considered that the "maturation of information storage" during this period is such that auditory behavior is subject to environmental modification. Consistent with this conclusion is the observation of Bridger (1961) that auditory frequency discrimination may be observed in a two day old infant. This same phenomenon was pointed out more recently by DiLeo (1967) who claims that certain Russian investigators have "demonstrated the neonate's ability to discriminate sounds of different frequency . . ." This very early and apparent maturation of auditory discrimination ability is what Murphy (1969) called "the phenomenon of selectivity." He observed ". . . the normal infant at the third week after birth ceases to respond to the sounds consistently present in the environment while responding often with startle or distress to uncommon sounds even though these are presented at significantly lower intensities."

Some concern needs to be expressed as to whether auditory maturation specifically can really be distinguished from neurosensory maturation generally. This question becomes critical in the case of the infant who appears not to respond. This matter was reviewed by Reichstein and Rosenstein in 1964. Downs (1962) considered that the hearing impaired child responds like the normal child if the sound is loud enough for him to hear and also if no other central nervous system involvements exist, but Fisch (1964) felt that tests of responses of newborns would not be confused by brain damage which did not involve the auditory pathways. If they do, of course, considerable confusion may result. Kimball (1967) discussed peripheral and central auditory disorders and pointed out that one could easily be mistaken for the other in the testing of infants. One must exercise care, however, in assuming either that peripheral or central disturbance may cause a lack of response. Buch (1966) in his study of the temporal bone of the neonate concluded: "The results militate against a central lesion being the only pathological basis." He was confident of the possible presence of loss of peripheral function. The issue may be, what does one expect of auditory maturation at birth? DiLeo (1967) suggested "One cannot properly speak of perception at birth as that would mean that a stimuli [sic] could be interpreted, and that is impossible without prior experience. Reception is more appropriately applied to the newborn's sensory function." The matter of reception versus perception may also be considered under the heading of cortical intervention in responses to acoustic stimuli. This topic is taken up in a later section. However, it is for this difficulty that William Hardy (1967) felt constrained to point out: "Inasmuch as hearing is a learned function which takes months of appropriate stimulus and response, the idea of trying to demonstrate deafness immediately after birth seems somewhat presumptuous." The consensus seems to be, however, not that we are attempting to demonstrate deafness or its absence immediately after birth, but are attempting to demonstrate instead that the neurosensory functioning of the newborn is intact. Such functioning includes, but is not limited to, auditory receptivity. Certainly it seems to be generally concluded, with some very notable exceptions, that the auditory function is sufficiently mature at birth or before such that its status can be accurately observed. Most workers in the area would probably agree with Eisenberg's (1970) observation that ". . . the human newborn emerges as a thoroughly remarkable creature. He is perceptually mature in a great many ways and better organized than casual inspection would suggest. His auditory behavior, which seems to depend on both central and sensory phenomena, does follow 'rules'. . ."

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* Although the famous Dr. Spock (1969) told us that "A newborn baby seems to be deaf the first day or two because of fluid in his inner [sic] ear."

## STIMULI

Perhaps the issue which needs to be discussed is not whether the neonate can respond but what are the stimuli which would cause him to respond. I raised this identical question in my 1969 paper: "What stimuli will cause an infant to behave in a way that would permit observers to determine if he responds to sound?" Eisenberg (1965a) showed that selected acoustic events have differential functional properties that are innate, unconditioned, and very probably relevant to the ontogeny of communicative functions. She has considered stimulus variables repeatedly. In 1967 she was concerned with stimulus significance; in 1969 with functional properties of sound and concluded: ". . . that the average newborn discriminates sound on the basis of parameter variables and the organization of the stimulus envelope."

What stimuli have been employed for auditory screening of neonates? If we look first at the frequency parameter, there has been a general preference for frequencies around 3,000 Hz. This was apparently concluded by 1967 when Rudmose said: "It is generally accepted that the critical frequency range for screening the hearing of the neonate is centered around 3,000 Hz." Downs and Sterritt (1964) used a narrow band of noise which peaked at 3,000 Hz. Later observations tend to concur with the conclusion of Heron and Jacobs (1968) to the effect that "A warbling note as opposed to a pure tone is necessary to evoke responses." Actually, as early as 1934 Stubbs had observed that the response activity of the neonate is proportional to pitch up to 4,096 Hz. Thompson in 1962 believed that stimuli of low pitch would tend to inhibit activity. Eisenberg (1969) concluded that neonates respond preferentially to the frequencies found in language but employ a "differential coding mechanism" for high frequencies. Earlier Eisenberg, Coursin, and Rupp (1966) had employed a stimulus descending continuously in frequency from 5,000 to 200 Hz in four seconds and repeated every ten seconds. They observed that this was a useful stimulus for indicating differences among neonates. Many investigators, however, have preferred rather broader spectrum noises. The many brilliant studies of the group at Johns Hopkins University (e.g., Hardy, Dougherty and Hardy, 1959) used a "clacker". You will recall that Marquis (1931) used a buzzer. The most interesting and relevant stimulus was employed by Roberts and Campbell (1967): the sound of human heartbeats. In general, however, there has been a demonstrated preference for higher frequencies usually around 3,000 Hz. Recently, however, Ling, Ling, and Doehring

(1970) were unable to find stimuli of 3,000 Hz any more effective than stimuli of 2,000 Hz. Nevertheless, there does seem to be some general agreement on the necessity for these tones to be warbled. The instrument described by Rudmose in 1967 uses a pure tone of 3,000 ± 150 Hz, varying 40 times per second.

In all clinical audiology the parameter which is of greatest interest is intensity. The basic question asked in clinical audiology is: how quiet a sound can the patient hear? This is a serious problem in the matter of testing neonates because, for the most part, neonates seem to be just uninterested in quiet sounds. It is for this reason, as pointed out above, that we can really only hope to discriminate by mass screening techniques those infants who are deaf from those who are not deaf. That is to say, we can hope to identify only those infants who fail to respond even to loud stimuli. Haller (1932) observed that disturbance increases with intensity. Stubbs (1934), on the other hand, observed that response activity increases irregularly with increases in intensity. Eisenberg (1969) considered the intensive responses to be very significant as she observed that "Intensity-bound behavior, as measured by threshold and loudness functions, has exact correlates in later life." But consistent with the observation that neonates respond only to loud stimuli Rudmose (1967) observed that repeatable reactions from newborns require stimuli on the order of 80 to 90 dB to demonstrate normal hearing. In the next section we consider the nature of responses to various stimuli but in general it may be observed that stimuli under 50 or 55 dB Sensation Level do not usually elicit responses in the newborn.

Not only are frequency and intensity parameters of consequence defining what constitutes a stimulus for a neonate, but also temporal factors are of concern. One of the things which seems to favor eliciting responses has to do with the rise time of the stimulus. Thompson (1962), for example, pointed out that ". . . abrupt auditory stimuli tend to evoke greater infant motility. . ." Many observations have been made as to what sort of durations constitute abrupt stimuli. As early as 1934, Pratt observed that an auditory stimulus repeated only once every ten seconds arouses a more clearly defined response than when the same stimulus is repeated every 60 seconds. This suggests the obvious possibility of habituation during silent intervals. We will return to this subject later. The comment with respect to the abruptness of the stimulus was found in the advice given by Herer (1967): ". . . observe the first stimulus administered because, frequently, repetition of the stimulus is not helpful and often confuses the picture." On the other hand, a signal to which the infant had adapted did not seem to become abrupt when some property of it was changed (Leventhal and Lipsitt, 1964). Adaptation to a repeated stimulus was effective, and when changes were introduced they failed to produce dishabituation. The habituation of certain electrophysiological responses to auditory stimuli has repeatedly been shown and is discussed in the appropriate section. For the moment we may cite the 1968 investigation of Clifton, Graham and Hatton in which it was observed that there was no habituation of heart rate responses of newborns for stimuli up to a duration of two seconds but there was when the stimuli exceeded that duration. It appears, however, that the directly measured physiological responses may be more sensitive to duration than are directly observed behavior responses. For example, Stubbs (1934) found that the response activity is proportional to duration up to 15 seconds. The cardiac response, as we have observed, becomes inversely proportional to duration when the duration exceeds two seconds.

Eisenberg (1969) attached considerable significance to the durational properties of stimuli which elicit certain responses. She said: "Pattern-bound behavior is uniquely selected, and it may reflect fairly high-level eighth nerve mechanisms."

It may be seen, in summary, that the parameters of frequency, intensity, and duration all contribute independently as well as interactively with the responsivity of the neonate. He seems to be most sensitive to frequencies in the neighborhood of 3,000 Hz, but recent data suggest that the boundaries of that neighborhood might be rather far from the center. Moreover, he seems to be especially sensitive, as are we, to loud noises; and relatively insensitive to quiet ones. And he seems to react to those stimuli which have some surprise value in time; and also those stimuli which are of relatively brief duration.

## RESPONSE PATTERNS: BEHAVIORAL

The caption on this section "Response Patterns: Behavioral" and the caption on the section which follows "Response Patterns: Electrophysiological" were not intended to suggest that behavioral responses are other than physiological. It would be foolish to suggest that the behavior of an organism is in any way independent of its physiology. The apparent dichotomy suggested in the organization of these two sections is only to point out that there are some responses which may be observed by looking at the infant and there are other responses which may be observed by looking at some sort of recording of the infant. It is the first class of responses which we consider in this section.

Birns et al. (1965) observed that human neonates are affected differentially by various auditory stimuli. Such an observation is consonant with that of Thompson (1962), to which we referred earlier, ". . . abrupt auditory stimuli tend to evoke greater infant motility; whereas continuous auditory stimuli of low pitch and moderate intensity tend to inhibit activity." There are two responses which have been repeatedly noted in the neonate. These are the eye blink response (auro-palpebral reflex or cochleo-palpebral reflex) and the Moro reflex. Downs and Sterritt (1964), for example, observed both eye blinks and Moro reflexes. Quite a variety of responses was observed by Eisenberg et al. (1964) including motor reflexes, eye reflexes, arousal, and orienting-quiet responses.

It appears that the response most frequently and reliably observed has been the auro-palpebral reflex. As early as 1946 Froeschels and Beebe observed that the auro-palpebral reflex was the most common response. Hahlbrock (1959; 1962) has intensively investigated the auro-palpebral reflex. Undoubtedly the most famous and perhaps the most significant study of the auro-palpebral reflex was that of Fröding (1960). Fröding subjected 2,000 newborn infants up to 30 minutes of age to acoustic investigation in an attempt to produce an easy and simple test. He discovered the auro-palpebral reflex to be the most reliable of any response. The reliability and significance of the auro-palpebral response were discussed by Wedenberg in 1963 (a). He concluded from his results that such hearing tests are extremely reliable (1963b). Herer (1967) observed that eye blink responses "can be elicited as early as the first 24 hours of life by a moderately loud stimulus." The auro-palpebral reflex has also been discussed by House, Linthicum and Johnson (1964), Mahler and Wagner (1967), and by Newby (1964) in his popular text. The nature of the auro-palpebral response was described by Taft and Cohen (1967) as: "The cochleo-palpebral reflex, in which a loud noise produces a blink and sometimes a startle reaction, is another example of the elicitation of the blink response by a stimulation of an adjacent organ, in this case the auditory system. The presence of this reflex, the only consistent auditory reflex found in newborn infants, is dependent upon a normal auditory conduction and neuronal system . . ."

There is a certain amount of controversy, however, as to whether or not the auro-palpebral reflex is the *only* consistent neonatal response. Many authorities have considered the Moro reflex to be also a consistent neonatal response. For example, Darley (1961) observed startle responses (i.e., Moro reflexes) to sudden noises. Frisina (1963) taught us that neonatal responses "are essentially all-or-none." He included the Moro reflex among the "overt readily observable responses . . ." And again in his widely read text, Newby (1964) said: "The response of a newborn infant . . . is a startle or Moro's reflex . . ." And lest one think the Moro response is unique to American or European infants, it should be pointed out that it was also discussed by Maezawa (1965) in Japan. There is considerable doubt, however, that the Moro response is a genuine auditory response in any part of the world. Taft and Cohen (1967) suggest the following interpretation:

> The primary stimulus responsible for producing the Moro reflex has not been clearly defined. The reflex was at one time considered an auditory phenomenon since it could be elicited by banging on the top of a table upon which an infant was resting. However, under similar testing conditions the identical stimulus applied to a hard-surfaced immobile table rarely produces a Moro response. Instead the Moro reflex occurs as the result of any sudden movement of the head and neck causing retroflection.

Essentially the same observation was made by Parmelee in 1964 to the effect that neck movement seems to be essential for a significant Moro reflex. In general, then, the Moro reflex ought not be taken as a reliable neonatal response.

Another response which is sometimes easy to observe has been mentioned a few times in the literature. This response takes the form of an alteration of respiratory behavior. Suzuki, Kamijo, and Kiuchi (1964) considered sudden deep inspiration among the most reliable responses of neonates to pure tone stimuli. Mahler and Wagner (1967) evaluated what they called breathing-test audiometry in the awake neonate. The most intensive and extensive investigations of respiratory responses have been those undertaken in South Africa by Heron and Jacobs (e.g., 1968; 1969). Their observations are consistent with those of Suzuki that a most common response is a sudden inspiration or a gasp. They observed the gasp occurring at three respirations after the cessation of stimuli.

Other basic responses have also been observed upon occasion. For example, Relke and Frey (1966) investigated the middle ear reflex of the newborn. As early as 1930, Pratt, Nelson, and Sun observed the incidence of various behavioral responses to acoustic stimulation. They noted that 35 percent of the responses were movements of extremities, 34 percent movements of eyes, 26 percent general body movement, 3 percent vocal responses and 1 percent facial movements. In general one must accept the conclusion of Martin and Stendler (1953): "The neonate . . . will modify his responses to sound according to the duration and intensity of the stimuli."

In general, that behavioral response upon which most of us have relied has been the auro-palpebral response. It is this response which has been noted repeatedly by the most astute observers (e.g., Wedenberg, 1963). While many researchers have attempted to employ the Moro reflex as an indicator of auditory intactness, this reflex is in serious doubt for this purpose. Other responses such as alterations in respiratory behavior are just beginning to be investigated. If I may interject a personal opinion at this point, it is my belief that those behavioral responses which are most valid are those elicited in the absence of an observer (cf., Jazbi, 1968; Butterfield and Hodgson, 1969).

## RESPONSE PATTERNS: ELECTROPHYSIOLOGICAL

The difference implied by separating electrophysiological responses from behavioral responses centers solely on what it is that one observes. In the case of behavioral responses the observer looks at the infant. In the case of what we call here electrophysiological responses the observer attaches an instrument to the infant and then watches the instrument. Some of these methods have been labeled objective audiometry (for example, by Beagley and Knight, 1966). One somehow doubts that electrophysiological methods are necessarily more objective than behavioral methods as we have defined them. In the case of behavioral methods one looks at the infant and makes a subjective decision. In the case of electrophysiological methods one looks at a piece of paper or an oscilloscope or some other display and makes a subjective decision. It seems to us that one is not more or less objective than the other. It is simply a matter of what it is one can be objective about. The term objective was also used, for example, by Long, Hilger, and Roth in 1967.

Lowell (1967) listed the electrophysiologic methods which have been used as hearing tests with infants. Among them were Galvanic skin response, cochlear potentiometry, plethysmography, computer averaged evoked cortical responses, and pupillometry. Similary, Jacobs and Heron (1970) have published data on recordings of respiration, eye movements, and skin galvanometry.

The most widely used electrophysiological method of audiometry has been the averaged evoked encephalic response. It is not the purpose of this review to discuss in detail the great many studies which have been done on this fascinating topic. The reader is referred to the outstanding collection of papers edited by Donchin and Lindsley (1969). Similarly, one of our most renowned scholars, Dr. Hallowell Davis, has done extensive and intensive research on this topic and has published many papers (e.g., Davis and Onishi, 1969). It is not our purpose here to review the entire subject but to point to some significant studies with specific reference to the neonate.

To avoid confusion we propose a symbology to avoid having to use such a long term as electroencephalographic audiometry or evoked encephalic audiometry or evoked response audiometry, etc. It has become popular to refer to this process as EAR, evoked auditory response. I find this term unsatisfying, because it does not specify what the response is. Let us then use the symbology ERA and mean by that encephalic response audiometry, and then later we can turn our attention to CRA, cardiac response audiometry. There seems to be some issue whether the encephalic response can be reliably elicited in the newborn. Callaway (1969) pointed out: "Evoked potentials show dramatic changes with maturation. The auditory evoked potential is obtained easily in infants . . . but it does not show the marked changes with maturation that are found in the visual evoked potential . . ." Davis and Onishi (1969) concluded: ". . . that the V potential shows no measurable maturation." In the same study though, they suggested: "Unfortunately the effects of sleep complicate the study of V potentials in infancy. The normal state of the neonate is the sleeping state . . ." Nevertheless, Callaway (1969) observed ". . . good AEP can be obtained during sleep . . . It has no measurable effect on the thresholds of infants. This is fortunate because it is difficult to find a time when a small infant is quiet except when he is asleep." There seems to be a difference of opinion here as to whether or not it is desirable for the neonate to be asleep during encephalic audiometry. One opinion says it is difficult to find the neonate when he is not asleep and that sleep affects the response. Another expert says while it is difficult to find a neonate awake, sleep has no significant effect on the encephalic response. A study by McCandless (1967), which included infants both within and beyond the neonate stage, concluded that the responses were 79 percent satisfactory. The lack of complete satisfaction, and the difficulty in eliciting responses even in the normal infant, was observed in a paper by Rapin and Graziani (1967) which observed that the proportion of false negatives was larger under sedation than during sleep. The stage of sleep also seems to matter. The study of Weitzman, Fishbein, and Graziani (1965) found the amplitude and latencies associated with the encephalic auditory response to be significantly smaller in active sleep than in quiet sleep. In a later study Weitzman, Graziani and Duhamel (1967) observed the effect of maturation and behavioral state of prematurely born infants tested during the neonatal period and at one, three and six months after discharge from the hospital. The effects of maturation on these infants were clear in the encephalic auditory response. It has been shown repeatedly, however, that CRA can be used for the diagnosis of severe hearing loss in infancy (Barnet and Goodwin, 1965; Barnet and Lodge, 1966, 1967). In the last of these three papers it was concluded: ". . . that EEG audiometry may be used in the diagnosis of hearing loss in the infant regardless of etiology." On the other hand, McCandless (1967) felt considerable difficulty in the interpretation of ERA as changes in the alertness of the infant influence the response waveform. He felt only experienced persons ought to be using this technique.

Especially in light of some of the difficulties and confusion surrounding encephalic response audiometry many authorities have turned their attention to cardiac response audiometry (CRA). One of the more extensive researchers in this area has been A. K. Bartoshuk (1962a, 1962b). He was among the first to observe that the nature of the neonatal cardiac response to acoustic stimulation was in the form of acceleration and that this response could be habituated and dishabituated (1962a). In that study, he examined 60 neonates, 30 of whom received a stimulus every six seconds and 30 of whom received a stimulus every 60 seconds, both receiving stimuli over a total period of 16 minutes. The group receiving the more frequent stimulation showed greater response decrement (habituation) across all trials. However, for all 60 infants an increase in intensity level of the stimuli led to dishabituation. He observed a similar phenomenon with respect to the frequency parameter so that if he changed the direction of frequency change, dishabituation occurs. In another study (Bartoshuk, 1962b) he observed that cardiac acceleration was a reliable response and its reliability increased with age over the first four days of life. He thought at that time that habituation might represent some form of learning rather than some form of neural fatigue. In that same year Beadle and Crowell (1962) determined that ". . . no consistently discernible pattern of response was evident." They noted the ". . . dependence of post-stimulus heart rate on prestimulus heart rate . . ." In 1964, Bartoshuk determined reliably the linear relationship between cardiac acceleration and stimulus intensity. This linear relationship appears upon a logarithmic plot. In 1965 Bridger, Birns, and Blank were able to compare behavioral ratings with heart rate measurements. In an early member of a series of studies Keen, Chase, and Graham (1965) observed that a habituated heart rate response would be retained by a neonate for 24 hours and that the habituation was a function of stimulus duration. More specifically: "The heart-rate response of newborns to auditory stimulation was found to be an inverted U-function of stimulus duration" (Clifton, Graham and Hatton, 1968). Again they observed that the cardiac response to acoustic stimulation took the form of acceleration. In fact, the accelerative response ". . . returned to, but not below, prestimulus levels" (Graham, Clifton, and Hatton, 1968). And, in a still later study from the same group, Clifton and Meyers (1969) showed that the nature of the cardiac response changes by the age of four months: "While the newborns had responded to this auditory stimulus with cardiac acceleration, the older infants showed deceleration." It appears, therefore, that the evidence of maturation may be stronger in CRA than in ERA. Again, Steinschneider, Lipton and Richmond (1966) observed the relation of the cardiac rate response to sound level. Meyers and Gullickson (1967) also observed the relationship of the evoked heart rate response to the acoustic pattern. A general evaluation of the cardiac response method of audiometry was stated by Jasienska et al. (1967) to the effect that the ECG as a method of "aurocardial reflex" is objective and sensitive. Some of us (and I advisedly say us) are leaning more towards CRA in preference to ERA on the grounds that it may be a more accessible response. There is also the possibility that it may be a more legitimate response of the peripheral auditory mechanism. Moreau, Birch, and Turkewitz (1970) observed that autonomic responses habituate with greater difficulty than do musculoskeletal responses. The matter of cortical mediation in autonomic (e.g., cardiac) responses is still open. It remains for anencephalic infants to be examined by these respective methods.

It might be disturbing to some, as indeed it should be, that both ERA and CRA are not measurements directly of the activity of the ear. Such methods may be possible, however, as evidenced by a recent paper and film by Aran (1970) and an earlier publication of Bordley, Ruben, and Lieberman (1964). Aran (1970) referred to this procedure as "electro-cochleography" and Bordley, Ruben, and Lieberman (1964) referred more directly to what they were actually doing (i.e., the acquisition of cochlear potentials in human beings). Bordley, Ruben, and Lieberman were not dealing with neonates but they observed that some persons who might be considered deaf by the more customary tests were indeed observed to have functioning cochleas to the extent that they demonstrated cochlear potentials. Aran recorded "click-evoked cochlear responses" in children as young as a few days of age. He describes this as "electrophysiological study of the peripheral receptor." And this is exactly the point: it would be desirable for us to be able to separate responses of the peripheral receptor from the activity of the central processor. ERA probably is unable to do so. CRA data remain equivocal on this point, although one is tempted to say that CRA responses are autonomic in nature. Clearly, however, electro-cochleography measures the periphery directly. It may eventually be the case that both electro-cochleography and either (or both) ERA and CRA will be required to distinguish between peripheral and central auditory disabilities.

The reader may be struck by the lack of mention of the Galvanic Skin Response (GSR) or of electro-dermal response audiometry (ERA, again). It is our impression that the literature has rejected the electro-dermal response as a valid response in neonates or, perhaps, even in older children or adults.

## ENVIRONMENTAL FACTORS

Virtually everyone who has published on this subject has referred to the apparent importance of the environment in which the neonate is tested. No one, however, apparently has discussed this matter in detail. There is a startling paucity of information on the influence of environmental factors upon the responsivity of the neonate to acoustic stimulation. In her several papers on the subject Rita Eisenberg (e.g., 1966) has referred to variation in the environment as influencing the responsivity of the newborn. She has noted that there is a significant difference between constant stimuli and pattern-bound behavior. That is to say, the neonate responds in one way to those stimuli which are constantly present in his environment and to those stimuli which are presented in a determined pattern as, for example, by an experimenter or observer. It has been suggested, however (Caziarc, 1963), that such examinations can be conducted in the pediatrician's office. Griffiths (1967) recommended free field testing in a sound room. McCroskey (1967) recommended hospital screening. Miller (1966) did a follow-up study and concluded that testing in a well baby clinic was a valid procedure. But none of these authors discussed environmental factors specifically. Finally, Fisch (1965a) proposed using recorded sounds for infant audiometry. It was startling to the present author in organizing the bibliography for the purposes of writing this review that there were no cards filed under the title "Environmental Factors." It appears that no one whose work has been brought to or come to our attention has considered environmental factors specifically. Perhaps this indicates the area of our greatest need in the matter of neonatal screening.

## INFANT STATUS AT THE TIME OF TESTING

Infant status for the purposes of the following discussion is interpreted in both of two ways. The first has to do with the status of the infant with respect to his gestational age; that is, the significance of prematurity versus full term delivery. Secondly, the other and more obvious interpretation of infant status has to do with the condition of the infant at the moment of tests. Both of these items are discussed in the literature, but one finds a relatively small number of studies devoted primarily to these topics. Fisch (1967) has called attention to both the "state" of the child and to the manner in which the examiner handles the baby. He also in that paper referred to external conditions. This is perhaps the only paper with these topics as its primary subjects.

The topic of the differences among premature and full term infants has already been discussed under the heading "Maturation." One needs only to be reminded in this context that to the extent that maturity (or prematurity) may be interpreted as the state of the infant it influences his responsivity. Recall that Eisenberg, Coursin, and Rupp (1966) were able to discriminate among newborns assigned in three different categories on the basis of risk. They observed that high risk subjects did not habituate to acoustic stimulation; suspects took twice as long as apparently normal infants and high risk subjects did not habituate to stimulation at all. In a paper by Field et al. (1967) prematures showed a decreased number of responses to the lesser of the two stimulus levels with respect to the responsivity of full term, well born infants. Of interest to the present writer was a recent paper by Sanders, Friedman, and Weintraub (1970) dealing with the effects of incubation on neonates. The incubator has long been a "bogey man" in the study of the auditory responsivity of the premature neonate. Sanders, Friedman, and Weintraub observed that the noise levels generated within incubators in common use are below the level considered hazardous for adults. However, one does not know if the criteria for hazardous noise exposure apply equally to neonates. But there is another very important and interesting problem raised by these authors: ". . . prolonged exposure to this undifferentiated acoustic environment may have an adverse effect on the early stages of auditory perceptual development in this group of infants who represent a high risk for neuropsychiatric sequelae." This seems to be a most important observation, one which has been chronically overlooked in our studies of the premature neonate. For a long time we have suspected that prematurity in and of itself may lead to auditory deficits. More recently we have suspected that the noise levels generated inside the incubators may lead to auditory deficits in premature neonates. The wisdom of Sanders, Friedman, and Weintraub is that neither of these may be the problem but instead the problem may be one of sensory deprivation (or distraction) during a critical period for the development of auditory perception. This seems to me to be a most important topic with reference both to environmental factors and to the status of the infant at the time of testing.

What about the well, full term infant? What influence does his status at the time of testing have upon his responsivity? Eisenberg et al. (1965) observed that differences in activity state exert significant effects upon auditory behavior. Fisch (1965c) referred to children's "ability to listen" rather than their ability to hear. And Bench (1970) has directed our professional attention again to the Law of Initial Value. Lewis, Bartels, and Goldberg (1967) have suggested the influence of the state of the infant upon his heart rate response to tactile stimulation. Again, Fisch (1970) has called attention not only to the type of stimulus but also to "the circumstances of its application." Bench (1969a) has shown that neonatal responses are elicited "by modifying this state of arousal . . ." Still further Bench (1969b) observed that neonates two to six days old quieted more readily to auditory stimulation than when they were unstimulated. Similarly, Birns et al. (1965) observed inhibition caused by acoustic stimulation in the "aroused" human neonate. Frisina (1963) cited: ". . . diminished activity in presence of bell sounds." Jazbi (1968) has been concerned with observing the activity of the infant in his crib from a remote location. It has seemed to a number of persons, in fact, that the state of the infant may be influenced by the presence of an observer. It may be wise, therefore, to remove the observer from the neonate's immediate environment and to provide him with a remote observation condition.

## BEHAVIOR OF OBSERVERS

The behavior of the observer of the neonate, as suggested in the concluding comment in the previous section, may be critical. Downs (1967b) has observed "the audiologist's inquiry starts with the very beginnings of deafness – at birth – and proceeds to continued identification of hearing status following birth." The nature of the inquiry, however, may or may not require the physical presence of the audiologist. Downs and Sterritt (1964), as we mentioned earlier, found very high interobserver reliability; which means that one observer saw what another observer saw. We also pointed out that reliability does not necessarily imply validity. There is no a priori reason to believe that, while the observers saw the same behavior, that behavior was in response to acoustic stimulation. It was noted recently, for example, by Ling, Ling, and Doehring (1970) ". . . many of the observed responses to sound appear to be of doubtful validity . . . Observers are likely to accept behavioral changes arising for other reasons as positive responses." Therefore, observer variables may be critical in interpreting the results of tests (Ling, 1970). The behavior of observers has not been monitored with sufficient care for us to fully appreciate its influence upon the responsivity of the neonate or upon one's interpretation of his response. In a very recent paper Hamilton, Derbyshire, and Joseph (1970) noted the interaction of the observer with the infant and pointed out that the observer is indeed part of the total test situation. This subject has not been adequately investigated except by Ling, Ling, and Doehring (1970).

## FOLLOW-UP STUDIES

How do we know that what we do is of any benefit? Very few persons, in fact, have done longitudinal studies or have attempted to follow up both those infants who did and did not respond at the time of birth. The most extensive work has been that reported by Downs and Hemenway (1969) on the screening of 17,000 newborn infants. Their data showed the incidence of hearing loss at birth to be one in 1,000, a figure somewhat higher than that noted in many other studies. In comparing these infants at the age of five months with normal hearing children of the same age, differences could be observed in vocalization. It seems that the content of the vocalization of hearing impaired infants is comparable with that of normal infants but its quantity is lacking. Rechecks, however, reveal a number of errors. In assessing the errors Downs (1967b) noted: "A fairly high rate of false positives is found in the programs: one and one-half percent of the total population failed to respond to the test signal and 97 percent of these are ultimately cleared." But those remaining children apparently are positively identified with considerable accuracy. Redell and Calvert (1969) found that "A follow-up of all children who 'failed' the testing indicates that newborns in other than a 'high risk' category were identified as having hearing loss." This was also the principal finding of the now classic study of Fröding (1960). Of the thousands of infants he examined within the first half hour of life all of those who survived and who did not respond upon recheck were later found to be deaf. What about those who are not deaf? Wedenberg (1963b) retested the normal children of his 1956 study when they were five years of age. All but one of these children were found to have normal hearing. He found the results of the first hearing test were verified in all but two of his cases.

It seems, therefore, that in the hands of competent, astute observers neonatal screening tests may be valid and reliable. The problem which faces us for the immediate future is how to generate either competent, astute observers or techniques or to develop techniques to make the rest of us appear to be competent and astute.

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This section contains all the items identified for this review, and is not limited to those items actually cited in the preceding. Therefore, it is not only a list of references but is a bibliography. The style of referencing and the abbreviations used for journals are, in the main, those of *DSH Abstracts*. Others conform to our idea of what they would be in that publication.

We have not seen everything listed herein, and may have included some items which are inappropriate. Also, we have undoubtedly (if unintentionally) omitted some important items; for this we apologize both to our readers and to the authors of omitted works.

Part I contains all items we have located on neonatal audition and peripheral items which have been cited in the text. Part II consists of those items which deal with infants beyond the neonatal period, or which deal with neonates in areas other than hearing. We decided to include Part II as we had located the items in the process of finding the more immediately relevant ones.

The "we" used in the above is to reflect the considerable assistance of my graduate assistant, Miss Mary Goldstein. We are also appreciative of the several partial bibliographies we received from many colleagues.

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## PART II

This section contains those items which (a) deal with neonates but not their auditory behavior, (b) deal with the auditory behavior of infants beyond the neonatal period, or (c) could not be identified. It is likely that we have made more mistakes and done injustice to more authors in this part than in Part I. However, since these items were revealed in the various bibliographies we have studied, it seemed well to include them here.

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# SCREENING PROGRAMS FOR THE DETECTION OF DEAFNESS IN NEWBORN INFANTS – A NEONATOLOGIST'S OVERVIEW

Sheldon B. Korones, M.D.*

The deep interest of the American Academy of Pediatrics in the problem before us is signified by its delegation of two liaison members to the Joint Committee on Newborn Hearing, which also includes representatives of the American Speech and Hearing Association and the American Academy of Ophthalmology and Otolaryngology. This committee, led by Mrs. Marion Downs, was formed because concern with early identification of impaired hearing was common to its three parent organizations. It has functioned with noteworthy cohesiveness; the diverse professional origins of its members having proved to be its principal strength because of pervasive intellectual honesty, a mutual deference to the respective purviews of its constituent specialties and an unfailing awareness of the common goal. I cite these attributes because they seem to characterize the spirit with which this conference was organized.

A nationwide interest in the identification of neonatal hearing impairment has been generated recently by audiologists, and the possibility of such early detection has intrigued and challenged the pediatrician. Quite frankly, it also may have stirred a few of us because evaluation of neonatal auditory function is not ordinarily included in the long list of items with which we are preoccupied. We have had similar experiences in the past. Recall, for instance, that approximately 20 years ago the ophthalmologists directed our attention to retrolental fibroplasia; and having been passed the torch, we pursued the problem vigorously. A short time later the role of oxygen therapy and the iatrogenic nature of the disorder were clearly demonstrated in a collaborative study involving pediatricians and ophthalmologists at several institutions. Examination of neonatal optic fundi has since become a standard procedure in certain circumstances. Perhaps you have a similar torch to pass. The possibility of early detection of deafness by methods which are scientifically sound, elicits a receptive response from those of us concerned with the intact survival of neonates.

One wonders why the nursery is considered the best place to screen infants for a hearing defect. Though we are concerned with the earliest possible detection of deafness, what is lost by postponing the procedure for a few weeks or months: At present, the most compelling argument for nursery screening seems to be one of logistics, for at no other time or place can so many young infants be tested so easily. We should realize however, that most of our small communities cannot mount effective screening programs and that a large segment of the neonatal population would not be reached unless regional centers performed the procedure; and this would have to transpire after the infant has been discharged. Nevertheless, the nursery does offer the best available opportunity for testing large numbers of infants.

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Several methodologic difficulties inherent in current screening techniques have been cited recently, and they are mentioned in the statement on newborn hearing screening which was produced by the Joint Committee and published by its parent organizations. As a consequence, the problem of false positive and false negative results arises and this is always a basic consideration in any mass screening program. The supposed behavioral responses to sound stimuli may be more apparent than real; differentiation between spontaneous activity and a true response is difficult. Thus theoretically, a baby could "pass" in error. Of even greater concern to the neonatologist are those infants whose failure to respond is spurious, and who are thus categorized as deaf, or possibly deaf, by whatever terminology is applied to their status. The resultant stress upon maternal-infant relationships is impossible to calculate but would nevertheless be quite real. Consider further the untested high-risk baby whose condition has produced considerable family distress, and has also necessitated a protracted period of separation from his mother, which by itself impairs the evolution of normal maternal attitudes. Our response to this type of situation has recently led to an "open door" policy in the high-risk nursery wherein mothers are invited, at the earliest possible postpartum date, to fondle their babies and to participate in caretaking procedures such as bathing, feeding and dressing. In the face of this effort to normalize maternal-infant interaction, one hesitates to complicate matters by offering even a tentative pronouncement in regard to questionable hearing status; particularly if it is predicated upon methods that are still controversial.

Perhaps the quest for deaf neonates does not require as its initial act, the presentation of acoustic stimuli to all babies in the nursery. If detectable neonatal deafness is a rare occurrence in the absence of a suggestive history or other demonstrable abnormalities, would it be anachronistic to suggest that we begin the screening process with a history and physical examination? If the latter is being performed satisfactorily by physicians, the identification of deafness as suggested by the demonstration of other abnormalities is not. The stimulus to pursue this problem must come from those most sensitively concerned with it, and those of you who are old enough to understand will be nostalgic about my request to "pass the ammunition."

Of more direct interest to this audience is the possibility of utilizing historic data to anticipate the likelihood of congenital deafness in an infant at risk. I have heard Mrs. Downs expound upon the merits of a high-risk registry and have been impressed with its potential value as a primary screening device. If a soundly devised questionnaire can be shown to effectively preselect infants in need of hearing evaluation, the testing itself could be accomplished in a leisurely fashion and in greater depth by those most qualified to do so. These preliminary inquiries could be applied universally because their execution would not require specialized qualifications of participating personnel. A larger segment of the newborn population would thus be scrutinized. I hope this approach receives some consideration during our workshop sessions.

It seems important to assess the influences, if there are any, of several neonatal pathophysiologic states on the behavioral response to sound. The clinically obvious ones are not a problem; they can simply be avoided. I refer to a few abnormalities which are either extremely subtle in their clinical manifestations or are not at all perceptible. For instance, can we expect accurate interpretation of results when testing a low birthweight infant who is not in a thermoneutral environment. Among the responses to cold stress, hyperactivity is the one that is of particular interest to us here; spurious results are likely to be frequent. Asymptomatic hypoglycemia is not uncommon among infants who are small for gestational age, usually occurring between 24 and 72 hours after birth. We should know if such infants respond to testing in an identical fashion as normoglycemic controls. The same questions can be posed for hypocalcemic infants and for those premature babies who develop metabolic acidosis during their second or third weeks of life. These are a few of the issues which should be resolved eventually if neuromuscular responses to sound stimuli are to be utilized as indications of normal hearing.

There are 3.5 million babies delivered in this country annually and 2,000 or 3,000 of them are deaf at birth. Identification of these infants during the neonatal period or soon thereafter would constitute the first important step in alleviating the difficulties that confront them. A universally applicable screening procedure is feasible, but its validity must be documented before its widespread use, lest we create more distress among our patients and their families than already exists.

# NEUROSENSORY FACTORS IN NEWBORN HEARING

Louis Gluck, M.D.*

*Reliable* statistics of school children with hearing handicaps are not readily available, estimates varying between 1 and 21 percent. The lower figure (1 percent) represents findings of the Health Examination Survey of 1963-65, analyzed and reported by the National Center for Health Statistics. Children 6-11 years of age were tested for pure tone loss with an estimated 213,000 children in this 6-11 year range who are handicapped. No reliable figures are available which estimates how many infants are affected.

Any compromise in normal communicative processes may produce major social, behavioral and widespread physiological complications. The younger the affected individual so handicapped the more all-engulfing becomes the loss and the more imperative it becomes to identify hearing losses as early as possible, hopefully in the newborn period.

Traditionally, hearing disorders are divided into three categories by location and defect:

1. *Conductive*, where sound transmission through the external and middle ears are affected.
2. *Sensorineural*, where the properties of hair cells as transducers and of the peripheral nerve as conductor are affected.
3. *Central*, where the actual processing is affected of the auditory signals and other auditory information in higher centers of the central nervous system.

To understand the origins and functional aspects of handicaps, it is helpful to consider the embryological development of the ear.

## EMBRYOLOGICAL ORIGINS

### *Primordial Tissues*

The three major areas, external, middle, and inner ears begin from separate areas in the embryo. All three basic primordial tissues, ectoderm, mesoderm and endoderm are involved.

Continued cell division after fertilization results in differentiation of the morula into a trophoblastic layer of cells, a trophoblastic cavity and an *inner cell mass* which will develop into the embryo proper. A second cavity, the amniotic sac, forms above the inner cell mass. By eight days of fetal life the inner cell mass differentiates into

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*ectoderm*, in contact with the amniotic cavity, and *endoderm*, in contact with the trophoblastic cavity. Some "primary" mesoderm then appears between the ectoderm and endoderm. The embryonic cells divide, forming notochord and then neural groove. At the caudal end of the notochord, the primitive streak forms, elongates, and in its proliferation, seeds mesodermal primitive streak cells, "secondary" mesoderm, between ecto- and endodermal layers.

The ectoderm gives rise to:
- External ear
- Outer tympanic membrane
- Membranous labyrinth
- Organ of Corti
- Semicircular canals
- Utricle
- Saccule

The endoderm forms:
- Inner tympanic membrane
- Eustachian tube

The mesoderm differentiates into:
- Ossicles
- Middle layer of the tympanic membrane
- Mastoid and petrous temporal bones

### *Pharyngeal Derivatives*

By three weeks, growth of the neural tube and of the anterior foregut is marked. Five pharyngeal pouches form, with arches and grooves. The first arch becomes the mandible, the second arch the hyoid bone. The external auditory meatus results from the first cleft, the "cervical sinus" from the second. Clefts or grooves demark one pouch from another. The cervical sinus houses arches 3, 4, and 5 and is the seat of branchial clefts and fistulae.

With growth, the first cleft invaginates. Inside, part of the second pouch extends anteriorly and laterally, toward the first cleft. An ectodermal plug separates this extension, the *tubotympanic recess* from the exterior. The layer of ectodermal cells adjacent to the tubotympanic recess together with its own outer layer of cells form the tympanic membrane. The tubotympanic recess will become the eustachian tube.

### *Auditory Vesicle*

The otic (auditory) placode appears above the first external groove opposite the hind brain. At first this is a thickening of ectoderm. This grows and invaginates into a pit which then is closed over by the surface ectoderm and becomes the auditory vesicle or otocyst, surrounded by mesoderm. With growth, the tubotympanic recess intervenes between the otocyst and the external auditory meatal area, establishing the ultimate gross relations of the three parts of the ear.

The auditory vesicle elongates. On the medial surface, at five weeks, a tubular diverticulum becomes demarcated. This will continue to differentiate into endolymphatic sac and duct. The larger portion of the chamber (utriculo-saccule), forms into semicircular canals and cochlear duct at six weeks. By nine weeks, differentiation has occurred so that the disc-like canals have become complete semicircles. The cochlear duct continues to coil, a process complete by 12 weeks. The auditory vesicle meanwhile, at eight weeks, changes into a cartilaginous capsule. The scale tympani and scale vestibuli extend in this cartilage along the sides of the cochlear duct until they fuse at the tip forming the helicotrema. Ossification of the cartilage begins and completes between 16 and 23 weeks.

Thus, the otic vesicle actually is the primitive membranous labyrinth while the mesoderm in which the otic vesicle lies differentiates to form the bony labyrinth surrounding the membranous labyrinth.

### *Middle Ear*

In the formation of the middle ear, the tubotympanic recess, an endodermal tissue, forms the lining membrane, eustachian tube and lower tympanic membranes. The ossicles form from adjacent mesoderm in which at 12 weeks, they are embedded. The mesoderm decreases in cellularity and loosens. By 20 weeks the ossicles lie in a connective tissue matrix. By 22 weeks the tubotympanic recess expands. The lining epithelial membrane invades and wraps around the ossicles and forms the middle ear air space in a pneumatization process.

### *Internal Ear*

The internal ear, lying in the petrous portion of the temporal bone, is in two parts: The bony or osseous labyrinth and the membranous labyrinth. The bony labyrinth is relatively much larger than the membranous labyrinth which is surrounded by fluid in the perilymphatic space. In the part of the bony labyrinth that houses the organs of balance, the perilymphatic space is filled with a mucus-like fluid supported by a fine fibrous network. The perilymph is much thinner and there is no fibrous network in the bony labyrinth surrounding the organ of hearing.

### *Bony Labyrinth*

The structure of the bony labyrinth is divided into three areas:

The *vestibule* is the central area, joining the five openings of the semicircular canals and the scala vestibula of the cochlea.

The *semicircular canals* form the posterior bony labyrinth and contain semicircular membranous ducts that occupy perhaps one-fourth of the space. Each canal has an ampulla at one end, a dilatation containing a vestibular sensory nerve ending.

The *cochlea*, the anterior bony labyrinth, is a tube that forms a spiral. It is divided into two parts along the entire length of the tube by a crest of bone together with a basilar membrane. The two halves of the tube communicate at the tip, the helicotrema.

*The Cochlea*

Nerves for the 1¾ turn of the cochlear tube go through a perforated base into the cone. They pair off successively toward the spiral lamina where the nerves enlarge, forming a chain of ganglia in the root of the osseous spiral lamina. The length of the cochlear tube is about 30 mm.

The membranous cochlear duct is a tube within a tube with the scala tympani, the perilymphatic space surrounding the cochlear duct. The membranous cochlear duct is also known as the scala media. The cochlear aqueduct connects the scala tympani with the subarachnoid space. The part of the membranous cochlear duct epithelium which rests on the basilar membrane is differentiated into the organ of Corti.

On section, the cochlear duct inside the bony cochlea actually is triangular. One side, the short side abuts the bony cochlea; the middle side, with the organ of Corti sits on the basilar membrane, and the long side, the vestibular membrane of Reissner, separates the cochlear duct from the perilymphatic space.

*The Organ of Corti*

The basilar membrane is fibrous and is said to contain about 24,000 fibers in each human ear. The epithelium which lies upon this basilar membrane is divided into two zones, the inner *organ of Corti* and the outer cells of Claudius. Supporting cells and hair cells make up the organ of Corti. The supporting cells have a variety of structural arrangements. They form a triangular space, the tunnel of Corti, on the inner (medial) side. The inner rod cells of this space support one row of hair cells. On the lateral side of the tunnel, there are three or four rows of hair cells alternating with other structural cells, the supporting cells of Deiter. The cells of Deiter sit on the basilar membrane and support a hair cell between each adjacent pair.

Laterally are other supporting cells, the cells of Henson and the cells of Claudius in particular.

Each hair cell is a columnar cell and has from 20 to 400 hairs on its outer surface. There are about 3,500 inner hair cells and about 12,000 outer hair cells. A tectorial membrane emerges from the osseous spiral lamina to overlie and make contact with the hairs from the hair cells in a gelatinous matrix.

*The Neurological Connections*

The auditory nerve (cochlear and vestibular branches) emerges from the internal auditory meatus and enters the brain stem between pons and medulla. The cochlear nerve forms from the central processes of the nerve cells that make up the spiral ganglia of the cochlea. The peripheral processes connect with the organ of Corti.

In the brain stem the cochlear nerve divides into two branches; one enters the ventral cochlear nucleus in front of the inferior cerebellar peduncle; the other branch passes laterally and enters the dorsal cochlear nucleus behind the peduncle.

The ventral cochlear nucleus fibers end in the dorsal nucleus of the trapezoid body of the same or opposite side. The second neuron fibers of the opposite side behave similarly and the two sets of fibers of the opposite side behave similarly and the two sets of fibers from the trapezoid body, from which another neuron system emerges as the lateral lamniscus. The lateral leminsci on each side contain neuron fibers connected with all four nuclei.

In the midbrain some fibers of the lateral lemnisci end in the inferior quadrigeminal body; others go on to the medial geniculate body where final hearing neurons form the acoustic radiation, passing laterally below the lentiform nucleus to the acoustic cortex.

The acoustic cortex is in the superior temporal gyrus, immediately below the lateral sulcus on the lateral aspect of the cerebral hemisphere. Hearing from each ear is relayed almost equally to the acoustic cortex of each side. Thus for deafness in either ear to arise from central causes the acoustic paths from both ears must be impaired.

### *External Ear*

As mentioned previously, the dorsal end of the first branchial cleft is the primordium of the external auditory meatus. It is medial to the mesoderm separating it from the tubotympanic recess. A solid plug of ectodermal cells persists until week 28 when it canalizes to form the external meatus.

In forming the external ear proper, six tubercles appear at six weeks gestation, at the first cleft; three tubercles on the mandibular arch and three on the hyoid arch. By 12 weeks there is fusion of all tubercles except the ventral mandibular one. This is accomplished by a general second arch mesodermal growth.

### *Mastoid*

Pneumatization of the mastoid process will not be covered in this discussion except to point out that it begins in the few weeks prior to birth and is completed normally at the end of the second year of life. By six months of age there is x-ray evidence of air cells adjoining the antrum.

## PHYSIOLOGICAL CONSIDERATIONS

### *Matching Impedances*

In the conduction of sound – for those with electronics background – a natural impedance mismatching must be overcome between air and fluid. Sound waves traveling in air must transmit their wave motion to the fluid medium bathing the organ of Corti. The sound resistance of fluid allows only some 0.1 percent of the energy in an air wave to enter; the remaining energy gets reflected from the surface. Thus matching air and water impedance requires an acoustic transformer.

### *Transforming Sound Pressure*

One hundred years ago Helmholtz suggested that sound pressure is transformed by a lever action of the ossicles and a hydraulic action due to differences in surface areas of tympanic membrane and stapedial footplate. The ossicular leverage develops around an axis running through the anterior process of the malleus and the short process of the incus. The relative difference in length between the handle of the malleus and the long process of the incus contributes the mechanical advantage (about 2-2.5). The tympanic membrane attaches to the stapes as a piston so that a hydraulic action results. The force is exerted over the whole tympanic membrane focused on to the footplate of the stapes with a gain in pressure/unit area. Mechanical advantage from the ratio of the two areas is about 20.

### *Cochlear Stimulation*

Although sound normally enters the cochlea by the oval window it may enter also by the round window and conducted to the cochlea via the bones of the skull. The cochlear capsule vibrates, the inertia of the contents allowing a *lag* to develop between movements of hair cells and capsule walls that results in stimulation. This lag provides a differential movement and the sound stimulus. Precisely how the energies of sound with the multiplicity of frequencies and intensities is abstracted by the cochlea and transmitted to the cortical auditory centers is not entirely clear.

Activation of the stapes initiates a mass action with movement of the entire fluid as the oval window transmits vibration instantly from drum to stapes. The basilar membrane assists in the transfer of motion by vibration.

In order for the quick adjustment to be able to take place it probably is necessary to involve the principle suggested by Bekesy of the traveling wave, which travels from one end of the cochlea to another, reaching maximum amplitude at some point by vibration of the basilar membrane causing an eddy current in the cochlear fluid which actually stimulates hair cells.

When the basilar membrane is stimulated at very low (e.g. 60 cycles/second or below) frequencies, the entire membrane vibrates. As the frequency rises, and definitely above 4,000 cps, relatively great selectivity takes place in the membrane and only portions seem to vibrate, possibly reflecting the place of maximum amplitude and not that of definite selectivity.

### *Electrical Potentials in Cochlea*

Several types of electrical potentials can be picked up following acoustic stimulation. Two types of potentials from the cochlea, one abolished by hypoxia and the other obtainable for a long period after death. Auditory nerve action potentiates (spike discharge); and changes in the position resting potential of the scala media. Stimulation of various places on the basement membrane results in potentials in various cortical areas.

## THE NEWBORN IN JEOPARDY

In the examination of the newborn infant, the physician is required to exercise observational skills that are far beyond those required in the evaluation of any other single group of patients. In relation to hearing incapacities there are a number of conditions that should alert the physician that an infant is or very likely may be hard of hearing. An infant at risk may be a representative of a wide variety of stresses.

### *Maternal Factors*

*Maternal disease or chemical inhibition* may cause damage to the fetus with resultant hearing loss or frank deafness. *Maternal rubella* is the best known and most common single cause of sensorineural deafness. Associated with a variety of other malformations, the infant most commonly presents with petechiae which may be large "blueberry muffin" spots, low platelets, congenital heart disease, cataracts, mental retardation, osteomyelitis, large liver and spleen, and low birth weight. Other maternal infections that may be associated with deafness in the offspring include Asian influenza, infectious mononucleosis, and other viral infections sporadically. Syphilis in the mother may cause deafness in about 5 percent of the offspring. Whereas the viral diseases mentioned may be associated with defects of organogenesis, luetic infections seem to be destructive lesions. Syphilis is a relatively late infection in the fetus since the spirochetes seem unable to cross the placenta before about the 20th week of gestation.

Maternal disease associated with an increased incidence of deafness importantly includes diabetes mellitus. This disease is well known to be associated with an incidence of malformations significantly higher than normal.

### *Ingestion of Drugs by Mother*

Almost any drug can cross the placenta. Many drugs taken by the mother preferentially have higher concentrations in the fetus than in the mother. A large number of relatively common drugs the mother may ingest either electively or prescribed by a physician may cause deafness, particularly if taken in the first trimester of pregnancy. These include: salicylates, quinine, a variety of antibiotics that are ototoxic (streptomycins, neomycin, kanamycin) various aniline dyes, carbon disulfide, carbon monoxide, and, of course, thalidomide.

### *Birth Injury*

In the course of being born, a variety of accidents can beset the fetus, resulting in *anoxic* damage. These may include compression of umbilical cord, prolonged delivery, abnormal pressures on head during delivery, maternal analgesics which may depress infant, and aspiration of fluids. Birth injury of any sort should be included with this group.

### *Hyperbilirubinemia*

Erythroblastosis from Rh or other incompatibility of blood groups between mother and fetus may cause deafness secondary to hyperbilirubinemia which follows hemolysis of red blood cells. The hyperbilirubinemia may produce kernicteric changes and deafness. Hyperbilirubinemia in the absence of demonstrable hemolytic disease also may produce kernicterus and deafness. Here other modifying conditions may make the infant susceptible, such as acidosis due to hypoxia.

### *Neonatal Infections*

Infants with infections in the neonatal period may be treated with ototoxic antibiotics (e.g., see above) and suffer permanent hearing damage. Certain infections themselves may result in a sequela of deafness including especially the meningitides. Other illnesses with resultant deafness include measles and mumps.

### *Noise*

Loud and sustained noise may be injurious. Questions have been raised whether the level of noise in an incubator is actually acceptable.

### *Prematurity*

Prematurity seems to be a major problem associated with a high incidence of deafness. Whether the resultant hearing difficulties are related to bad medical practice, is not known or whether the premature infant inherently has hearing damage. One unanswered problem in this regard is whether the large number of ototoxic drugs almost routinely given to the premature (e.g. kanamycin) may not be excreted due to inadequately developed kidney function and may produce hearing damage.

### *Genetic Disorder*

A wide range of genetic disorders are associated with total or partial loss of hearing. These include:

Treacher-Collins syndrome
First arch syndrome
Osteogenesis imperfecta
Cleft palate and lip
Trisomies (21-23, 13-15, 17-18)
Albinism
Cretinism
Hunter-Hurler syndrome
Waardenberg's syndrome
Pendred's syndrome (goiter with nerve deafness)

## EXAMINING THE NEWBORN

Certain clinical findings, including thumb anomalies, amputations, other cranial nerve problems, low set or abnormal ears, palatine abnormalities, eye anomalies, pigmentation defects, etc., should alert the physician to the need to screen the hearing of the infant.

During the first month of life absence of certain signs may signify a hearing impairment. For example, a normal infant responds to sounds by a variety of gross bodily reflex movements. If a handbell is rung, he ceases activity. He should show a startle response to a sharp clack from 3-6 feet away. He should be awakened or disturbed by any loud environmental noises. At the very least, a normal infant will show the *cochleopalpebral* reflex, in which a loud noise produces a blink and sometimes a startle reaction.

Although it is not in the purview of this discussion to compare the various hearing tests, it would seem most valid and reliable to employ *cortical audiometry*. Cortical responses to auditory stimulation recorded by a *computer-averaging technic* should be made routine in the newborn. The test takes only about 30 seconds and measures cortical evoked responses to a 1,200 cps stimulus. This works equally well on infants asleep or awake. In the meanwhile until this procedure becomes widely available, any of several *screening* procedures involving a noise stimulus to elicit a startle or blink should be done routinely on all newborns.

However, a firm recommendation should be made that cortical evoked potentials analyzed by computer-averaging technic be made available to all newborns and that infants be tested in the newborn period. Ideally, telephone computer lines from all hospitals delivering newborns should connect with one or more computer centers where almost instantaneous evaluations could be given. Since there may be diversion of response and subcortical responses to stimuli, the cortical evoked potential is the only present test which can assure that there is intactness of the entire auditory tract.

Why does one insist that testing be done in the newborn period? Because sound sensory deprivation from birth has such a devastating effect on the entire development of the infant and child.

Is this a valid time to test, in the newborn period? Indeed it is. The inner ear is the only organ in the body that by *mid term* in the fetus has reached full adult size and differentiation, occurring long before the fetus has become even a viable premature. Furthermore, there is no doubt that the fetus in utero can hear, as shown by cortical evoked potential with electroencephalogram.

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# HEREDITARY AND CONGENITAL FACTORS AFFECTING NEWBORN SENSORINEURAL HEARING

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The purpose of this paper is to review the hereditary and congenital factors affecting newborn sensorineural hearing, giving criteria for suspecting hearing loss in the newborn. These clinical criteria include the following: 1) evidence of any of the hereditary congenital deafness syndromes, 2) history of infection during pregnancy, 3) prenatal exposure to ototoxic drugs, 4) birth injury, and 5) neonatal hyperbilirubinemia.

## HEREDITARY CONGENITAL SEVERE DEAFNESS SYNDROME

The more than 60 types of hereditary deafness include 17 known types of congenital severe hereditary neural deafness syndromes (Konigsmark, 1969, 1971). These syndromes account for more than one-half of the cases of congenital deafness (Lindenov, 1945; Sank and Kallman, 1963). For those interested in hearing loss in the newborn, it is important to have some acquaintance with these syndromes because: 1) knowledge of a family history of congenital hearing loss will alert you to the possibility of deafness in the child, and 2) knowledge of the syndromes with their associated defects caused by the same gene involving other systems will alert you to the possible diagnosis.

These hereditary deafness syndromes can be distinguished from one another by the mode of transmission (dominant, recessive, or sex-linked), by the type of hearing loss (conductive or neural), and by involvement of other organs or systems by the gene causing the hearing loss. The congenital severe hereditary deafness syndromes can be divided into four groups, according to other systems involved (no associated abnormalities, associated with integumentary system disease, skeletal disease, and associated with involvement of other systems).

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## HEREDITARY CONGENITAL SEVERE DEAFNESS WITH NO ASSOCIATED ABNORMALITIES

The majority of the congenital hereditary deafness syndromes are in this group. The six types of hereditary deafness show severe hearing loss at birth, some even progressing after birth. Patients with these types of deafness are otherwise completely normal.

### Dominant Congenital Severe Deafness

In this type of deafness, as in all of the autosomal dominantly transmitted types, a single abnormal gene is responsible for the hearing loss. About one-half of the affected child's sibs are well as a parent of the child and half of the parent's sibs will have this abnormality, for the single abnormal gene, when present, will manifest itself. The hearing loss shows little variation from one affected person to another; all patients have a congenital bilateral severe neural hearing loss of from 60 to 100 dB. They are mute unless given special speech training. Other audiometric tests cannot be done for too little hearing remains for valid testing. Caloric vestibular testing generally shows normal responses.

To determine the incidence of hereditary congenital deafness, Sank (1969) surveyed deaf residents in New York State. She concluded that autosomal dominant genes accounted for about 10 percent of all congenital deafness. Also Fraser (1964), studying the causes of deafness in 2,300 children in special schools, concluded that 10 percent of the deafness in this group was due to dominant inheritance. Since there are about 31,000 students in schools for the deaf in the United States, about 3,000 of these students most likely have dominantly transmitted congenital hearing loss.

Other conditions such as prenatal rubella, meningitis, or viral infections should be considered in the differential diagnosis. In dominantly transmitted deafness a good family history is important, identifying all members of the family with hearing loss as well as the age of onset and severity of the hearing loss. Even if both parents are deaf, the hearing loss may be recessively transmitted.

Characteristics of this disease include: 1) autosomal dominant transmission, 2) congenital severe deafness in all affected family members, and 3) generally normal vestibular responses.

### Dominant Unilateral Deafness

Although most types of hereditary deafness are bilateral with both ears equally affected, several types including Waardenburg's and Treacher-Collins diseases affect the two ears unequally. Kindreds in which affected persons had unilateral or bilateral severe neural hearing loss with no other associated abnormalities were described by Smith (1939) and by Everberg (1960).

Otological examination was normal in all five cases examined by Smith. All of the affected persons were born deaf. Four were affected in the right ear and four in the left; in one person the side was unknown and in one person the hearing loss was bilateral. Audiometric results were not presented on these cases; it is possible that there may have been some hearing loss in the supposedly normal ears. Caloric vestibular tests on the affected persons studied by Smith were normal.

In Smith's family no cause for the hearing loss was found other than hereditary. The pedigree shows 11 affected persons in four generations. Involved were four of seven sibs, their mother, two maternal aunts and their maternal grandfather. In the family described by Everberg, there were five persons affected in three generations. The proband was congenitally deaf in his right ear only, while his two sisters had bilateral congenital deafness and his mother had right-sided congenital deafness. The pedigree suggests that this disease is transmitted by an autosomal dominant gene which shows variable penetrance, sometimes causing bilateral congenital deafness and sometimes only unilateral deafness.

Other factors that may cause unilateral deafness are mumps, meningitis, otitis media, head injury, or vascular disease. These types of unilateral deafness show a fairly rapid onset and are not congenital.

Characteristics of this disease include: 1) autosomal dominant transmission, 2) congenital moderate to severe unilateral or bilateral hearing loss, and 3) normal vestibular function.

### Recessive Congenital Severe Deafness

In this type of deafness both genes at a locus on the pair of chromosomes must be abnormal for the disease to become evident. Since one chromosome is furnished by each parent, the parents may be homozygous or heterozygous for this defect. Thus in general about one-quarter of the patient's sibs will be deaf, and their parents frequently are related.

Sank (1963) in a population survey and twin study of the deaf residents in New York State, concluded that about 40 percent of all cases with early total deafness is of autosomal recessive origin. Fraser (1971) studied 2,355 children in schools for the deaf. He concluded that from 23 to 30 percent were deaf because of autosomal recessive inheritance. These figures may vary from area to area depending on the level of medical care and the degree of consanguinity in the population.

All persons with recessive congenital severe deafness have a profound bilateral neural hearing loss, although some affected persons have a minimal amount of hearing remaining. Pure tone audiograms show an 80 to 100 dB loss in all frequencies with absent bone conduction. Too little residual hearing remains for other audiometric tests. Examination of the ears, external auditory canals, and tympanic membranes show no abnormalities.

On only a few affected persons are vestibular findings available. In a study of two kindreds in Pennsylvania, Mengel, Konigsmark, and McKusick (1969) found vestibular function to be normal in 12 affected persons subjected to caloric vestibular tests. In another family studied by us vestibular tests showed moderate to complete paresis of vestibular function. It is possible that this test may help separate one of the types of recessive congenital deafness from the others.

A large number of pedigrees have been presented showing recessive transmission of congenital severe deafness (Hopkins and Guilder, 1949). It is quite clear that there are several different genes producing the same phenotype of congenital deafness. Mengel et al. (1969) studied the congenitally deaf persons in the Amish and Mennonite communities of Pennsylvania. In one case an affected person from the Mennonite pedigree married a deaf person from the Amish pedigree. Their three children had normal hearing. Since each pedigree showed autosomal recessive inheritance, the genes causing these two kinds of deafness must be at different genetic loci; the children were heterozygotes for both types of deafness but phenotypically normal. Several other authors have presented pedigrees in which each of two deaf parents had autosomal recessive deafness, and children from the unions had normal hearing (Stevenson and Cheeseman, 1956). Fraser (1971), reviewing 3,500 persons with profound hearing loss of early onset, concluded that the total number of such genes represented in his series is rather low, possibly four or five, and that one of these genes is relatively common. Chung and Brown (1970) studying pedigrees at the Clarke School for the Deaf, estimated that there were five recessive genes responsible for the deafness.

In studying a child with congenital severe deafness, the various causes of this type of hearing loss, including prenatal rubella, kernicterus, birth injury, streptomycin or other intoxications, viral infections, and otitis media should be ruled out. A careful otological examination and hearing history should be taken to evaluate these possible causes of the hearing loss. A family history is also important, for quite frequently the parents attribute the hearing loss to some infection or trauma although the real cause may be hereditary.

When it is determined that the congenital severe deafness may be on an hereditary basis, a careful pedigree is necessary to determine whether the transmission is dominant, sex linked, or recessive. A thorough physical and neurological examination should be done to search for other possible anomalies associated with the deafness which may help to define the disease. To differentiate the congenital from early onset recessive deafness, a careful history concerning development of speech, and attention to the degree of development of the patient's speech will help in this differential diagnosis. With a history of some speech development, and where some evidence of facility with speech is present, the diagnosis of early onset recessive deafness may be considered.

In all cases it is important to do vestibular testing to determine the intactness of the vestibular system. This test may well help to separate out one of the types of recessive congenital severe deafness.

Characteristics of this syndrome, as we now know it, include: 1) autosomal recessive transmission, 2) congenital severe neural hearing loss, and 3) generally normal vestibular function.

## Recessive Early-Onset Neural Deafness

A syndrome characterized by a congenital and early childhood severe neural hearing loss has been described in several kindreds (Barr and Wedenberg, 1964). The ten affected persons we studied in a Mennonite kindred were found, by their relatives, to have had some hearing at birth, responding to sounds and sometimes learning to speak a few words (Mengel, Konigsmark, Berlin, and McKusick, 1967). Progressive hearing loss occurred fairly rapidly between the ages of one and one-half and six years with severe hearing loss in all affected persons by six years of age. One child was able to attend a public school for several years before hearing loss forced her to a school for the deaf. Audiograms on the ten available affected family members showed severe neural hearing loss from 60 to 100 dB in all frequencies. The two on whom a small increment sensitivity index (SISI) test was done had a positive SISI score (90 to 100 percent) bilaterally suggesting a cochlear origin of the hearing loss.

To document possible early hearing, we tested the speech of two affected persons. Sonograms taken from these two persons were compared to those made from a normal hearing person and those from a congenitally deaf person. Sonograms from the congenitally deaf speaker, trained in an oral school, showed absence of high frequency information and marked elongation of speech, as compared to the normal control. Sonograms from the two affected persons from this family showed an intermediate type of speech pattern, much closer to that of normal than to that of the congenitally deaf person. Skilled judges, evaluating the voice tape recording, concluded that both patients must have had good low tone hearing at least until their fifth or seventh years of life.

A medical history, otological examination, and hearing history showed no cause for the hearing loss in the Mennonite family other than heredity. Sixteen members of the family were affected in three generations. Males and females are affected in equal numbers. Irregular dominant inheritance is a possibility. However the proband's parents were known to have fairly normal hearing as well as the previous generations of the family. The pedigree is most compatible with autosomal recessive transmission; this mode is also most likely for the four affected sibs in the family described by Barr and Wedenberg.

In order to separate this disease from the congenital severe hearing losses, a detailed early hearing history and, in later years, a sonogram or speech analysis will help by indicating whether patients learned some speech during early childhood. This disease must also be differentiated from the early onset sex-linked type of hereditary deafness. The pedigree is essential in this differential diagnosis. In the later disease essentially only males are affected with transmission by their mothers.

Characteristics of this syndrome include: 1) early onset of severe neural hearing loss with essentially no hearing after five or six years of age, 2) normal vestibular function, and 3) autosomal recessive transmission.

### Sex-Linked Hereditary Deafness

Sex-linked transmission of a congenital hearing loss has been described in several families (McRae et al. 1969). In each of these families congenitally deaf males were found in several generations. These males were born of normal hearing mothers who had deaf brothers.

Audiograms on affected persons in each of the families show a bilateral 70 to 100 dB neural hearing loss involving all frequencies. No abnormalities have been described in the ears, external auditory canals, or ear drums. Vestibular tests have not been described in any of the affected persons.

Each of the pedigrees shows recessive transmission of this defect. In the family described by Dow and Poytner (1930) there were nine deaf males in four generations. Sataloff, Pastore, and Bloom (1955) studied four generations of a family in which six males were deaf mutes. All were born of normal mothers.

Fraser (1965a), studying 2,750 cases of profound childhood deafness, concluded that about 6.2 percent of the male cases were due to sex-linked recessive deafness.

Characteristics of this hearing loss include: 1) sex-linked mode of transmission, 2) congenital severe neural hearing loss, and 3) mental deficiency in some cases.

### Sex-Linked Early Onset Neural Deafness

Sex-linked transmission of an early onset neural hearing loss in a family was described by Mohr and Mageroy in 1960. All affected persons in this family had some hearing in early childhood, developing speech in the early years. Hearing loss stopped further speech development in childhood.

The 11-year-old proband learned to speak, but somewhat loudly. At about three years of age there was a decline in his speech until he was quite difficult to understand by 12 years of age. Audiometric tests were not described. His 12-year-old cousin had similar difficulties beginning to speak until three or four years of age when a regression of speech and hearing began. Other affected persons in this pedigree also began speaking with then deterioration during childhood. One boy was tested audiometrically when he was 13 years old. A hearing loss of over 80 dB in all frequencies was reported. Vestibular tests were not described in any of the affected persons.

The pedigree showed only males affected in four generations of this kindred. Transmission was X-linked.

This disease may be the same as the sex-linked congenital deafness. However, speech in all of the persons affected with the sex-linked congenital deafness did not develop, nor was there any history that speech had begun in persons affected with this disease. In contrast, affected persons in Mohr and Mageroy's kindred showed evidence of progression of the hearing loss at a very early age, after beginning speech was learned and before reaching school age. Thus this type of hearing loss is probably different from the congenital severe nonprogressive sex-linked hearing loss.

The causes of early childhood deafness such as meningitis, virus infection, and syphilis should be ruled out in the differential diagnosis. A careful history determining whether there is any evidence of progression of the hearing loss in childhood is important in separating the congenital severe deafness which is nonprogressive from the sex-linked early onset neural deafness which shows progression in childhood.

Characteristics of this type of hearing loss include: 1) sex-linked transmission and 2) some hearing in early childhood but essentially no hearing by school age.

## HEREDITARY CONGENITAL SEVERE DEAFNESS ASSOCIATED WITH INTEGUMENTARY SYSTEM DISEASE

In five of these six syndromes, skin or hair pigmentary change is the major visible abnormality, while the sixth syndrome shows skin thickening over joints. In all syndromes there is severe congenital deafness.

### Waardenburg's Disease

The various features of this disease were crystalized by Waardenburg in 1951. In studying 840 deaf mutes in five Dutch institutes for the deaf he found 12 cases of this disease. In the families of his patients he found numerous other aspects of the syndrome. The most characteristic features of patients with this disease are the widely spaced medial canthi and flat nasal root with confluent eyebrows. Frequently patients will have variably colored irides and a white forelock. At least one-half have some hearing loss.

The hearing loss is quite variable, ranging from no clinical deafness to severe congenital neural deafness. Waardenburg estimated that 14 percent of all deaf mutes in the Netherlands have this syndrome. DiGeorge, Olmsted, and Harley (1960), suggested that about 2.3 percent of the congenitally deaf have this disease and estimated that there are some 400 children in the United States schools for the deaf affected by this syndrome.

The most complete survey of vestibular function in Waardenburg's disease was presented by Marcus in 1968. Of 18 affected patients in a family, only one had completely normal vestibular function.

Patients with this syndrome have several prominent facial features, including widely spaced medial canthi producing what appears to be widely spaced eyes. However the inner pupillary distance remains normal. The broad root is found in about 75 percent of those affected while about one-half of the affected have confluent eyebrows over the nasal root.

Heterochromia irides was found in about 25 percent of the cases studied by Waardenburg. The patterns of pigmentary changes involving the iris are varied. While some patients may have the classical heterochromia iridium with one brown and one blue iris, some patients show brown sectors in a blue iris and blue sectors in a brown iris. Other patients may show brown or blue irises bilaterally (Goldberg, 1966).

A white forelock originating at the hairline in the middle of the forehead and continuing posteriorly is found in about 20 percent of the patients with this syndrome. The size varies from only a few hairs to a large forelock. Premature graying of the hair, eyebrows, and eyelashes has been noted in several kindreds. Waardenburg found prematurely gray hair in four kindreds and suggested that this may be equivalent to the appearance of the white forelock in some individuals.

Several types of skin pigmentary changes have been observed in affected persons. These range from areas of vitiligo to areas of depigmentation with patchy areas of pigmentation. Striking depigmentation of the skin was observed in a case described by Klein (1950). In several patients described by Fisch (1959) areas including the arms and face showed a patchy or freckled hyperpigmentation.

All cases of this syndrome presented in the literature show dominant transmission though there is striking variation in the degree of penetrance of the various traits.

Major characteristics of this syndrome include: 1) dominant transmission with variable penetrance, 2) lateral displacement of medial canthi and lacramal points in all affected, 3) broad high nasal root in about 75 percent of those affected, 4) vestibular hypofunction in about 75 percent of those affected, 5) congenital mild to severe unilateral or bilateral neural hearing loss in about one-half of those affected, 6) hyperplasia of medial eyebrows in about one-half of the affected, 7) heterochromia iridium and loss of pigment epithelium of optic fundus in about 25 percent of those affected, 8) white forelock in about 20 percent of those affected, 9) skin pigmentary changes including vitiligo and spotty hyperpigmentation in less than 10 percent of those affected, and 10) hare lip and cleft palate in less than 10 percent of those affected.

### Dominant Albinism and Congenital Deafness

A single family with this disease was described by Tietz (1963). He studied a newborn boy, his older brother, and their mother; all three were albino, deaf, and mute. Other members of the family were normal with no evidence of hearing loss or skin pigmentary changes.

The mother and her two affected children were deaf from birth and had developed no speech. Pure tone auditory tests showed a severe bilateral neural hearing loss. No vestibular tests were described.

Examination of the three affected persons showed albinism involving the entire body with the exception of the eyes. Irides were blue and optic fundi were normal in color. There were no visual difficulties or photophobia. Two skin biopsies taken from the mother showed no melanin. The hair was normal in amount except for hypoplasia of eyebrows with only a few sparse hairs representing the brows.

The family history shows that 14 members of the kindred in six generations had a combination of albinism and congenital hearing loss. Transmission was from parent to child; about one-half of the children of each affected parent were affected. Albinism and deafness were combined in all cases. Thus, this familial disease is autosomal dominant in transmission.

Characteristics of this disease include: 1) autosomal dominant transmission, 2) albinism of entire body with exception of the optic fundi and irides, 3) scanty eyebrows, and 4) congenital severe deafness.

### Leopard Syndrome

A syndrome described as the "leopard syndrome" by Gorlin, Anderson, and Blaw (1969) has components including: Lentigines, Electrocardiographic defects, Ocular hypertelorism, Pulmonary stenosis, Abnormalities of genitalia, Retardation of growth, and Deafness. It is transmitted by a dominant gene with variable penetrance, and shows a fair degree of variation from case to case.

There is a marked variation in the degree of hearing loss in different affected persons. In the families described by Matthews (1968) and by Walther, Polansky, and Grots (1966) no mention was made of hearing loss in any of the affected persons. The mother and daughter described by us (Capute et al. 1969) both had congenital severe hearing loss with very poor speech development. Because of the severity of hearing loss, special audiometric tests could not be done. Caloric tests in these two patients showed no abnormalities.

The most obvious findings in affected patients are the multiple lentigines involving face, trunk, and extremities. In the mother and daughter described by us, the skin was normal until about one year of age when freckle-like lesions appeared on the neck and thighs. These increased in size and numbers, spreading to involve the entire skin surface. Although concentrated on the upper trunk and face, some may appear on the scalp, palms, soles, and genitalia.

Cardiac abnormalities are noted in most cases on physical examination and include a systolic murmur, loudest over the pulmonary valve. Cardiac catherization done on a five-year-old boy, showed a slight increase in right ventricular pressure, and a minimal pulmonary stenosis was diagnosed (Matthews, 1968). Both the mother and the daughter we described had a mild pulmonic systolic murmur. Catherization studies were not done.

There is mild growth retardation. Some patients show a moderate ocular hypertelorism. Other anomalies found by Gorlin and Anderson include: pectus carinatum or excavatum, dorsal kyphosis, winging of scapulae, mandibular prognathism, hypogonadism, undescended testes, and late puberty.

In each of the kindreds transmission was by autosomal dominant mode. Whether, as suggested by Gorlin et al., these various families represent aspects of the same syndrome or whether these might be different diseases is not completely clear at present. Considering the range of manifestations in different affected persons, it is most likely that this is a single disease transmitted as an autosomal dominant with a variable penetrance.

Characteristics of the leopard syndrome include: 1) autosomal dominant transmission with variable penetrance, 2) lentigines developing after birth, 3) electrocardiographic defects including widening of the QRS complex and bundle branch block, 4) ocular hypertelorism, 5) pulmonary stenosis, 6) abnormalities of genitalia including hypogonadism, 7) retardation of growth, and 8) deafness, variable in degree.

## Hereditary Piebaldness and Congenital Deafness

A syndrome consisting of piebaldness and congenital deafness appeared in two of three Hopi brothers described by Woolf, Dolowitz, and Aldous (1965).

The two brothers were born deaf. Recent audiograms showed a 60 to 100 dB neural hearing loss bilaterally. Hearing in the normal sib and both parents were normal. The affected boys attended a school for the deaf. They had not learned to speak. Caloric vestibular tests were normal.

The boys, eight and 12 years old, showed a similar pattern of depigmentation. Although the major part of their bodies, including the back and legs, showed normal pigmentation, the entire head and hair were depigmented as well as a strip across the upper chest and over both arms. Within all of these depigmented areas were numerous small spots of hypopigmentation and hyperpigmentation. The irides were blue with a pattern of very fine clumps of pigment closely and uniformly spaced throughout the retina. Vision was normal. The boys had normal intelligence and were doing well in school.

Although the parents were Hopi Indians from the southwestern United States, no consanguinity was known. There was no family history of pigmentary defects or of hearing loss in either parents' family. Still it is most likely that this syndrome is hereditary and is transmitted by an autosomal recessive gene, although sex-linked transmission cannot be excluded.

Characteristics of this syndrome include: 1) probably autosomal recessive transmission, 2) pigmentary changes including depigmentation of head and portion of arms with hyperpigmented spots in depigmented areas, and 3) normal vestibular responses, and 4) congenital severe hearing loss.

## Sex-Linked Pigmentary Abnormalities and Congenital Deafness

A Jewish family containing 14 deaf mute males in three generations was described by Ziprkowski and coworkers (1962) and by Margolis (1962). Four of the affected were studied in detail and all showed similar clinical features.

All affected persons were congenitally deaf. Otological examination showed normal auricles, canals, and ear drums. Audiologic examination showed no response to air conduction at frequencies above 500 Hz. Caloric vestibular tests showed no response in three affected patients tested. A fourth tested patient showed moderate bilateral depression of vestibular response.

The skin changes involved the entire body and were characterized by large leopard-like spots of hypopigmentation and hyperpigmentation. Areas of the skin were sharply demarcated with fairly symmetrical distribution of pigmentary change. Achromic areas were whitish-pink, while browned or pleomorphic hyperpigmented areas were mottled with shades of color varying from a few mm to several cm in size. At birth the skin was albino except for areas of light pigmentation over the gluteal and scrotal areas. Pigmentation gradually increased involving particularly the arms, legs, buttocks, and face. Only a few spots appeared on the scalp. The hair remained completely white even when growing from pigmented areas. The remainder of the physical examination showed no abnormalities. The affected children had normal intelligence.

Fourteen cases of this syndrome occurred in males in a single kindred. Transmission is sex-linked.

Characteristics of this disease are: 1) sex-linked transmission, 2) congenital severe neural deafness, 3) pigmentary changes of skin beginning in infancy and characterized by large irregular spots of hypopigmentation and hyperpigmentation, and 4) depressed vestibular responses.

### Dominant Keratopachydermia, Digital Constrictions, and Deafness

Congenital deafness; hyperkeratosis involving the palms of the hands, soles of the feet, knees and elbows; and ring-like furrows developing on the fingers and toes were the major findings of this syndrome affecting four members of a kindred described by Nockemann (1961) and a single individual described by Drummond (1939). The four affected persons described by Nockemann and the patient presented by Drummond were deaf mutes. Vestibular tests were not described.

Each of the affected persons developed, beginning at about two years of age, thickening of the palmar and plantar skin followed by involvement of the elbows and knees. Rubbing produced thickenings elsewhere. When the affected persons were about five years of age ring-shaped furrows began to develop on the skin and soft tissue of the middle phalanx of all fingers and toes. These were severe enough to require digital amputation in several of the affected persons.

In the family described by Nockemann, the four affected members included a 20-year-old man, his mother, maternal uncle, and grandmother. The pedigree showed dominant transmission of this syndrome. No family history was presented by Drummond in his description of a single case.

Characteristics of this syndrome include: 1) autosomal dominant transmission, 2) congenital severe deafness, 3) hyperkeratosis involving palms, soles, elbows, and knees, and 4) ring-like constrictions of the soft tissue of the middle phalanges of the fingers and toes.

## HEREDITARY CONGENITAL SEVERE DEAFNESS ASSOCIATED WITH SKELETAL DISEASE

There are two syndromes in this category. In one there is absence of the tibia and in the other there is absence of some digits. Congenital deafness is a characteristic of both of the syndromes.

## Recessive Absence of Tibia and Deafness

In four of six sibs Carraro (1931) described a syndrome of congenital absence of one or both tibias and severe congenital hearing loss. Each of the four affected children was born deaf. No further audiometric testing and no vestibular tests were mentioned.

The affected sibs were normal physically except for their lower legs. Two sibs had marked shortening on one and mild shortening of the other lower leg, while the remaining two sibs had marked shortening of both lower legs. Roentgenograms of the lower legs in the four sibs showed similar findings. There was striking shortening or absence of tibias bilaterally. The fibulas were shortened and bowed, sometimes extending proximally across the knee joint to rest adjacent to the femur.

This syndrome involved four of six sibs. Both of the parents were normal, as were the remaining two sibs, with no history of hearing loss or boney deformities in either family. This disease appears to be transmitted by autosomal recessive mode.

This syndrome is characterized by: 1) autosomal recessive transmission, 2) congenital absence of one or both tibias and shortened malformed fibulas, and 3) severe congenital hearing loss.

## Recessive Split-Hand and Foot Syndrome

In 1963 Wildervanck described two brothers with hand and foot deformities; they were pupils at the Institute of Deaf at Groningen.

The tympanic membranes of the four and six-year-old brothers were normal. The younger boy had a 40 to 100 dB neural hearing loss, while his brother had a bilateral 60 to 100 dB neural hearing loss. A caloric vestibular test on the older boy showed marked depression of the vestibular response with minimal nystagmus produced by cold water.

The six-year-old boy had syndactyly of the third and fourth fingers of the right hand and an absence of phalanges of the middle finger of the left hand. On the right foot the phalanges of the second toe were absent with proximal syndactyly of the third and fourth toes. On the left foot the phalanges of the second and third toes were absent. His four-year-old brother had absent phalanges of the third finger of the right hand while the left hand was normal. On the right foot, the phalanges of the second toe were absent with syndactyly of the third and fourth toes. On the left the phalanges of the second toe were absent.

Roentgenograms of the hands and feet confirmed absent or syndactylous digits. No other boney abnormalities were noted. The boys were described as using their hands well with no disability.

This syndrome, appearing in two sibs from normal parents, appears to be recessive in transmission. Characteristics of this syndrome are: 1) autosomal recessive transmission, 2) hand and foot deformities, including absent phalanges of the finger or toe, and syndactyly of some remaining digits, 3) a congenital neural hearing loss of 40 to 100 dB, and 4) depressed vestibular responses.

## HEREDITARY CONGENITAL SEVERE DEAFNESS ASSOCIATED WITH OTHER ABNORMALITIES

In this category congenital deafness is associated with abnormality of the eye, thyroid, or heart.

### Recessive Retinitis Pigmentosa with Congenital Severe Deafness (Usher's Syndrome)

Affected persons are born deaf but otherwise appear normal. There occurs a progressive visual loss due to retinitis pigmentosa, leading finally to complete blindness in the second or third decades of life. Vernon's (1969) study showed that from five to 10 percent of the congenitally deaf have Usher's disease.

Affected persons are born deaf. Testing shows a severe neural hearing loss bilaterally. However Vernon found some variation in hearing loss among eight cases with Usher's disease. Two patients had no response to pure tone testing. The remaining six patients had mild to moderate preservation of the low frequencies from 125 to 500 Hz. Vestibular testing was done on six patients studied by Vernon. All of these were described as "defective", although the type of testing was not mentioned.

Vision through childhood is usually normal with the onset in adolescence of a slowly progressive visual loss, first noticed at night. The visual fields slowly constrict and visual acuity decreases. In the fourth to sixth decades of life only minimal vision may remain. Ophthalmological examination shows a slowly progressive retinitis pigmentosa. Optic discs become pale, arteries become narrowed, and cataracts may appear.

Despite the large number of cases of Usher's disease described in the literature, only a few sibships have been studied in detail. Usher (1914), in his long treatise on the inheritance of retinitis pigmentosa, described four sibships with affected persons having deaf mutism and retinitis pigmentosa. Parents in each of the families were normal. Vernon (1969), among eight cases of Usher's disease, found two pairs of sibs. This syndrome is transmitted by autosomal recessive mode.

Characteristics of this disease include: 1) autosomal recessive transmission, 2) congenital severe neural hearing loss, 3) slowly progressive visual loss due to retinitis pigmentosa, and 4) vestibular defect.

### Recessive Goiter and Deafness (Pendred's Disease)

In 1896, Pendred described an Irish family in which two sisters in a sibship of 10 were deaf mutes and had goiter. Since then many families with this syndrome have been described.

Affected persons are usually born severely deaf although variations in hearing loss occur. Audiometric testing shows a bilateral 40 to 100 dB neural hearing loss, more severe in the high frequencies. In a few cases hearing loss may be minimal and occasionally one ear may be relatively spared (Fraser, 1965b). Positive recruitment was found in two cases tested by Fraser. This suggests that the auditory defect is in the organ of Corti. In several cases skull x-rays showed a normal middle and inner ear.

Caloric vestibular tests generally show depressed vestibular function although some authors found normal vestibular responses in some of their cases (Von Harnack, Horst, and Lenz, 1961).

Diffuse goiter usually develops about the time of puberty, later becoming nodular. In most cases the goiter is prominent although it may be minimal. Histologically the thyroid shows numerous hyperplastic nodules with marked pleomorphism of the parenchymal cells. The picture resembles that of the thyroid from goitrous cretins. Patients with Pendred's disease are usually euthyroid, although some patients are mildly hypothyroid.

Fraser (1965b) estimated that the population prevalence of Pendred's disease at birth is about 7.5 per 100,000. Pendred's disease has been described in a large number of sibships by numerous workers. The parents in most cases are normal with no evidence of goiter or deafness. Inheritance is by autosomal recessive mode.

Patients with Pendred's disease have been subjected to a large number of operations for removal of the goiter. The goiter invariably returns, following continued stimulation by thyrotrophic stimulating hormone (T.S.H.). The goiter is best treated by exogenous hormone which then causes a decrease of production of T.S.H. and of thyroid stimulation. If started early enough, the goiter may regress.

The syndrome of endemic cretinism and deafness is common in restricted areas such as the Alps, Andes, and the Himalayas, where iodine is deficient. These patients show mental deficiency and physical deformities of cretins. The perchlorate test in these patients is negative or equivocal, in contrast to the positive perchlorate test in patients with Pendred's disease.

Characteristics of Pendred's disease include: 1) autosomal recessive transmission, 2) symmetrical, generally severe, congenital neural hearing loss, 3) a positive perchlorate discharge test, and 4) goiter developing in adolescence.

### Recessive Heart Diseases and Deafness (Jervell and Lange-Nielsen Disease)

A syndrome consisting of congenital deafness, prolonged QT interval with Stokes-Adams attacks and sudden death in four of seven sibs was described by Jervell and Lange-Nielsen (1957). Since then 13 additional cases have been described (Fraser, Froggatt, and James, 1964). All of the cases have bilateral congenital severe neural hearing loss.

Affected children have "fainting spells" with sudden lapses of consciousness beginning between infancy and 12 years of age, but usually between three and five years of age. Death occurs between three and 14 years of age in over one-half of the cases.

Electrocardiograms in all cases are abnormal with prolonged QT intervals of about 0.5 sec. (maximum normal is 0.4 sec.), and with large T waves. The QT prolongation may vary within and between persons. Death, when it occurs, takes place during a syncopal attack and is probably due to cardiac arrhythmia.

This syndrome is transmitted by recessive inheritance and accounts for less than 1 percent of children with severe deafness.

Patients with congenital deafness and a history of periods of unconsciousness, sometimes diagnosed as epilepsy, should have an EKG done in order to clarify the diagnosis. The major features of this disease are: 1) autosomal recessive transmission, 2) congenital severe neural deafness, 3) prolonged QT intervals on the electrocardiogram, and 4) recurrent Stokes-Adams attacks beginning in early childhood and frequently resulting in sudden death.

## CONGENITAL NEURAL DEAFNESS DUE TO INFECTION DURING PREGNANCY

Although infectious diseases such as bacterial or fungal meningitis, congenital syphilis, and mastoiditis can cause deafness in childhood, they do not cause congenital neural deafness. The major infectious diseases causing congenital deafness are congenital rubella, congenital cytomegalic inclusion disease, and congenital toxoplasmosis infection.

### Congenital Deafness Due to Prenatal Rubella

The incidence of infants affected with prenatal rubella increases with epidemics of rubella. In the recent epidemic of 1963 to 1965, there was an increase in the incidence of deafness due to prenatal rubella to about 10 percent of all those born deaf (Bordley and Hardy, 1969). Infants with congenital rubella have a variety of defects of varying severity, depending on the time during embryogenesis that the infection occurred. The major abnormalities in affected infants are heart disease (50 percent), hearing loss (50 percent), cataract or glaucoma (40 percent), psychomotor retardation (40 percent), and neonatal thrombocytopenia (Cooper and Krugman, 1967).

Hearing loss can result from infection during any part of the pregnancy. However, it is most frequent in children whose infection occurred during the first trimester, among whom 68 percent are deaf; infection in the second trimester causes deafness in about 40 percent (Bordley and Hardy, 1969). Borton and Stark (1970) presented the audiometric findings on 55 patients who had hearing loss due to rubella during the first trimester. Hearing loss was moderately severe to profound. There was no specific type of hearing loss, although 40 percent showed more severe loss in the higher frequencies. Most were neural, although about one-fourth had mixed hearing loss. The temporal bone histopathology shows varying degrees of cochlear duct and saccule involvement of the Scheibe type (Bordley and Alford, 1970).

It is important to recognize that about one-half of the mothers have subclinical rubella without a rash, giving births to infants with laboratory confirmed rubella (Bordley and Hardy, 1969), and that some patients with congenital rubella may be normal except for the hearing loss. Thus serological tests on the infant may be necessary to make the diagnosis.

Characteristics of this syndrome include: 1) prenatal rubella infection, 2) positive serology for rubella, 3) congenital neural hearing loss (50 percent of cases), 4) congenital cataracts, 5) congenital heart defects, 5) congenital miniaturization of fetus, 6) mental retardation, 7) mild brain damage, 8) delayed language development, and 9) thrombocytopenia.

## Congenital Deafness Due to Cytomegalic Inclusion Disease

Prenatal infection with the cytomegalovirus (CMV) causes a syndrome characterized by hepatosplenomegaly, jaundice, hemolytic anemia, thrombocytopenia, nervous system disease, chorioretinitis, and pneumonia. In some cases hearing loss is present (Emanuel and Kenny, 1966). Hanshaw (1970) estimated that about 4,000 children may be born each year with nervous system disease due to CMV infection. The diagnosis may not be apparent and requires identification of cytomegalovirus complement-fixing antibody in the serum.

Studies of temporal bones in two cases with CMV infection showed cytomegalic cells with inclusions in the inner epithelial layer of Meissner's membrane and stria vascularis with hydrops of the saccule (Myers and Stool, 1968; Davis, 1969).

Characteristics of this disease include: 1) positive CMV complement-fixing antibody in the serum, 2) hepatosplenomegaly, 3) hemolytic anemia and jaundice, 4) nervous system disease, 5) chorioretinitis, and 6) hearing loss.

## Deafness Associated with Congenital Toxoplasmosis Infection

Congenital toxoplasmosis, a protozoan infection causing hydrocephalus, chorioretinitis, and intracranial calcium deposits, has been implicated as causing deafness, although congenital hearing loss does not seem to be a prominent feature of this disease. Clinically affected infants have seizures, spasticity, and chorioretinitis. In 1961 Feinmesser and Landau described a 14-year-old boy with congenital toxoplasmosis and congenital neural deafness. However it is not clear that toxoplasmosis infection caused the deafness. Three of the four members of a family affected with toxoplasmosis described by Campbell and Clifton (1950), noted moderate progressive hearing loss beginning in childhood.

Kelemen (1958), in his study of the temporal bones of two infants with toxoplasmosis, described calcium deposits in the stria vascularis and spiral ligament as well as perivascular lymphocytes in the mesenchyme. These changes would be compatible with some degree of mixed hearing loss.

Characteristics of this disease include: 1) nervous system infection by Toxoplasma gondii with intracranial calcium deposits and hydrocephalus, 2) chorioretinitis, 3) positive Sabin test for toxoplasmosis, and 4) sometimes hearing loss.

## CONGENITAL DEAFNESS DUE TO OTOTOXIC DRUGS

Ototoxic drugs, taken during pregnancy, can result in congenital hearing loss in the infant. Some of these drugs include: streptomycin, dihydrostreptomycin, kanamycin, neomycin, ethacrynic acid, and thalidomide.

Although there have been several reports suggesting that ingestion of quinine during pregnancy causes congenital deafness in the fetus (Taylor, 1937), there is no increase in congenital deafness in malarious areas of the United States where quinine is taken by large numbers of persons (Winckel, 1948). Winckel concludes that congenital defects are no more numerous and no more serious whether the use of quinine has been excessive or moderate.

### Streptomycin and Dihydrostreptomycin

The ototoxic effects of streptomycin and dihydrostreptomycin are well documented (Heck, Hinshaw, and Parsons, 1963). Meurman and Hietalahti (1960) tested periodically the hearing of 389 patients treated for pulmonary tuberculosis in a sanitorium. They found that three and one-half percent developed hearing impairment, and they found that most patients tolerated high doses of streptomycin. However some were susceptible to smaller doses. They concluded that the hearing should be tested periodically, and that if slight hearing loss appears, medication should be changed. There are only a few articles describing fetal ear damage due to streptomycin. Conway and Birt (1967) found that children, probably affected by congenital exposure to streptomycin, may suffer from labyrinthine damage without hearing loss. Hearing loss involves high frequencies of about 8,000 Hz. Damage was not related to the fetal age when streptomycin was given or to the total dose. Both mother and child were likely to be affected. Among 17 children whose mothers received streptomycin during pregnancy, eight had slight abnormalities of hearing or labyrinthine function. Varpela, Hietalahti, and Aro (1969) studied the hearing of 40 children whose mothers had received streptomycin and/or dihydrostreptomycin at various stages of pregnancy. The hearing in all of these children was normal except in a single case who had a bilateral 60 to 90 dB high tone neural hearing loss. A caloric vestibular test showed no reaction from the right ear.

### Kanamycin

Kanamycin, in the adult, can produce hearing loss after administration of from 32 to 134 grams (Frost, Hawkins, and Daly, 1960). It is particularly toxic if there is renal insufficiency. It has a greater cochlear toxicity than dihydrostreptomycin but less than neomycin. No cases of hearing loss in infants following fetal exposure have been described. Sanders, Eliot, and Cramblett (1967) studied 20 children who received kanamycin in the neonatal period. These children all had normal neural hearing, though a few had conductive loss. However studies are needed on children who were congenitally exposed to this ototoxic drug.

### Neomycin

There are no reports of the production of congenital deafness by maternal administration of neomycin. However, it is clear that this antibiotic can cause deafness in adults. Lindsay, Proctor, and Work (1960) described a 50-year-old man who received 18 grams of neomycin over a 19 day period. Following this he was completely deaf. Histopathological examination of the inner ears showed an almost total loss of inner hair cells of the organ of corti. There was a loss of 60 to 100 percent of outer hair cells. Severe deafness can also occur following the administration of neomycin to an infant (King, 1962).

### Ethacrynic Acid

Ethacrynic acid, particularly on a background of poor renal function, can produce temporary or permanent neural hearing loss (Mathog and Klein, 1969). No congenital cases have been described.

### Thalidomide

In 1962 Lenz and Knapp directed attention to the possibility that thalidomide was responsible for the high number of children born with limb deformities. This was confirmed over the next several years. Ear abnormalities occurred when thalidomide was taken between the 35th and 45th day after the last menstruation. Ear deformities were combined with paralysis of the facial and oculomotor nerves in cases studied by d'Avignon and Barr (1964).

Usually the thalidomide induced hearing loss is conductive and due to atresia of the external auditory canal. However Rosendal (1963) described a thalidomide infant who died at four months of age. Autopsy showed a hypoplastic right petrous bone, while the left was normal. The internal auditory canals and the acoustic and vestibular nerves were absent bilaterally.

Fortunately thalidomide toxicity was never a problem in the United States, and is no longer available elsewhere.

### Summary

Although evidence is not overwhelming on the fetal effects of ototoxic drug ingestion during pregnancy, it is clear that these drugs cause hearing loss in children and adults. Thus in studying the congenitally deaf infant it is important to know if any ototoxic drugs were taken during pregnancy.

## CONGENITAL DEAFNESS ASSOCIATED WITH ANOXIC BIRTH INJURY OF THE BRAIN

Many reports have described hearing loss in cerebral palsy. However cerebral palsy may be caused by brain injury before birth, during birth, or shortly after birth. Thus it is difficult to know the effect of anoxic birth injury on hearing.

It is most likely that a fair percentage of children diagnosed as having athetoid cerebral palsy have anoxic birth injury, for anoxic birth injury affects the basal ganglia with status marmoratus causing athetosis. Some cases are due to neonatal hyperbilirubinemia. Hopkins et al. (1954) found in New Jersey that of 1,293 cases of cerebral palsy studied, 23 percent of those with athetoid cerebral palsy had hearing loss. Clear studies on hearing loss in children with anoxic birth injury are needed.

Hall (1964) found in 50 fatally asphyxiated infants, that there was a significant loss of neurons in the cochlear nuclei; he concluded that these nuclei were particularly susceptible to anoxia, and that this loss accounts for the hearing loss seen later in those who survive.

Characteristics of this syndrome include: 1) history of anoxic birth injury, 2) athetoid cerebral palsy, and 3) hearing loss in some cases.

## CONGENITAL DEAFNESS ASSOCIATED WITH NEONATAL HYPERBILIRUBINEMIA

If the bilirubin level in infants rises above 20 mg per 100 ml, the brain may be damaged by the toxic effects of bilirubin on neurons. In the neonatal period affected infants are jaundiced, apathetic, and restless. There may develop rigidity, spasms of the bulbar muscles, and seizures. Death may occur between the third and seventh days of life. Those infants who survive have sequelae including rigidity with involuntary movements of the trunk and extremities. In the mildest and more frequently seen form, the child develops chorea or athetosis at about two years of age; the gait is ataxic and there is emotional lability. Hearing loss, found in 20 to 40 percent of the cases, persists. Mental deficiency is usually present.

There is a wide variation in different studies on the percentage of affected children who have hearing loss. Cavanaugh (1954) and Keaster, Hyman, and Harris (1969) described an incidence of about 4 percent, while Crabtree and Gerrard (1950) found that 80 percent had hearing loss. The lower figures are probably closer to correct, for they are based on a more recent larger series of cases in which therapy was directed to keeping the bilirubin concentration below 20 mg per 100 ml. Keaster and coworkers studied 405 patients who had neonatal hyperbilirubinemia of at least 15 mg per 100 ml. Of these cases, 17 (4.2 percent) had mild to severe neural hearing loss. The two cases with severe hearing loss had bilirubin levels of about 17 and 22 mg per 100 ml, while the eight patients with moderate hearing loss had bilirubin levels of from 15 to 35 mg per 100 ml. The hearing loss was more severe in higher frequencies.

Fisch and Osborn (1954) found that of 891 children with congenital neural hearing loss, 27 (3 percent) had neonatal hyperbilirubinemia. The lesion causing this hearing loss is probably due to neuronal loss in the cochlear nuclei (Gerrard, 1952).

Characteristics of this syndrome include: 1) a history of hyperbilirubinemia (greater than 20 mg per 100 ml), 2) chorea or athetosis, and 3) hearing loss in some cases.

## SUMMARY

Hereditary and congenital factors are responsible for about 5,000 infants born each year with congenital severe deafness in the United States. These factors include genetic defects, maternal infection, prenatal exposure to ototoxic drugs, birth injury, and neonatal hyperbilirubinemia. The major clinical features of each of these types of congenital deafness are reviewed.

## ACKNOWLEDGEMENTS

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# HEREDITY AND CONGENITAL FACTORS AFFECTING NEWBORN CONDUCTIVE HEARING

Burton F. Jaffe, M.D.*

Conductive hearing losses in the newborn have not received as much attention as sensorineural hearing losses in the newborn. There are probably three reasons to explain the lack of emphasis on conductive hearing losses. The first is that the ears of newborns are not routinely examined in the newborn nursery and therefore certain diseases of the tympanic membrane and middle ear will not be diagnosed. Second, the hearing loss from a conductive disorder is more difficult for parents and nursery personnel to appreciate since the maximum conductive loss is only 60 decibels compared to the more obvious sensorineural hearing loss which can reach 110 decibels. Third, available audiometric testing of the newborn is not able to identify the mild to moderate hearing loss typical of a conductive loss, and therefore these lesions are not detectable on routine newborn screening tests.

The conductive hearing loss in the newborn may persist into infancy and early childhood and yet still be unidentified by parents or physicians. The sequelae of these hearing losses may include the late onset of speech, impaired speech, impaired language development, learning disabilities and perhaps even an altered behavior.

A sense of optimism should be associated with newborn conductive hearing losses since the hearing losses are frequently reversible. Otitis media in the newborn can be identified and treated by pediatricians and otolaryngologists. Structural deformities of the ear canal and middle ear ossicles may be suspected by the pediatrician and treated surgically by the otolaryngologist. In addition, amplification is available through the use of hearing aids. Certainly a potentially reversible lesion, as a conductive hearing loss, should attract our strong interest.

In general, the hereditary and congenital factors related to these conductive losses are not known, but where specific data is available they will be discussed.

I will emphasize four points. First, the tympanic membrane in the newborn can be examined. Second, the value of a pneumatic otoscopy to detect mobility of the tympanic membrane will be presented. Third, bilateral otitis media may occur in the newborn and certain high risk groups do exist for the development of otitis media. Fourth, structural deformities of the ear canal and middle ear ossicles can occur and the deformity may be an isolated ear anomaly or may occur in association with other anomalies.

**I. The Neonatal Ear Examination:** The newborn tympanic membrane can be examined as soon after delivery as desired and at any time until discharge from the newborn nursery. Vernix caseosum and blood may fill the ear canal initially but it quickly dehydrates in a few hours and becomes a mushy debris. After cleaning the ear canal, the tympanic membrane is visible. Although examination of the newborn tympanic membrane is not easy, it certainly can be learned and is a safe procedure.

A hand-held battery-operated otoscope with a closed examining head and a pneumatic bulb should be used. The use of a head mirror and reflected light in the newborn is far more difficult. A 2 mm. green nylon speculum is almost always correct to use for examining a newborn ear because it fits snugly into the ear canal and seals the canal for pneumatic otoscopy. Pneumatic compression of the tympanic membrane will cause the tympanic membrane to vibrate. Normally, the middle ear is filled with air so the tympanic membrane should vibrate briskly. However, when fluid exists in the middle ear, the tympanic membrane fails to move, moves slightly or moves laterally but not medially (inward).

Two instruments are useful in cleaning the vernix caseosum from the ear canal. A dull Buck curette #00 will fit through the otoscope head and attached speculum and is useful in cleaning out mushy or solid debris. A triangular applicator with a cotton twist at the end is useful in cleaning out moist debris.

The equipment for examination of the newborn tympanic membrane is illustrated in #1.

I prefer to examine the newborn with the aid of one assistant. To examine the left ear, the newborn is placed on its back, the physician stands at the head of the table, and the assistant stands at the right of the table. *(Illustration* #2) The assistant places her left hand on the newborn's head, turning it towards her and pushing it firmly against the table. *(Illustration* #3) Her right hand restrains the newborn's two hands and pulls them inferiorly so that the left shoulder will not rise to touch the left ear or the otoscope. No restraining sheets or wraps are used so that no time is lost in the examination. The physician stands at the head of the table and aims the otoscope towards the neck. This is valuable for two reasons. First, the ear canal does angle inferiorly, i.e. towards the neck, in the newborn. Second, the canal skin inferiorly is loosely attached and tends to obscure the view of the tympanic membrane. By aiming the tip of the otoscope inferiorly the skin can be lifted up by the speculum making examination easier.

It is important to pull the pinna inferiorly prior to inserting the speculum in order to stretch the loose skin of the neonate's ear canal. *(Illustration* #4) The ear canal in an adult or child is rigid because the inner two-thirds of the ear canal is tympanic bone which is not collapsible. However, in the newborn, the only bone in the ear canal is a ring of bone around the tympanic membrane. The rest of the ear canal is made of less rigid soft tissue and cartilage which allows the skin to collapse into the lumen.

With the otoscope held in the right hand, the speculum tip is then inserted into the left ear canal while pulling on the pinna with the left hand. *(Illustration* #5)

Cleaning the debris is then carried out through the otoscope. *(Illustration* #6) The otoscope is transferred to the left hand. The magnifying lens is positioned halfway to the side so that the curette held in the right hand can be placed through the otoscope

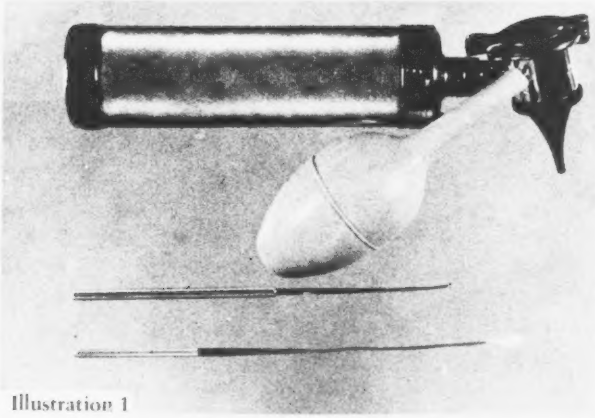
Illustration 1

Illustration 2

Illustration 3

Illustration 4

Illustration 5

Illlustration 6

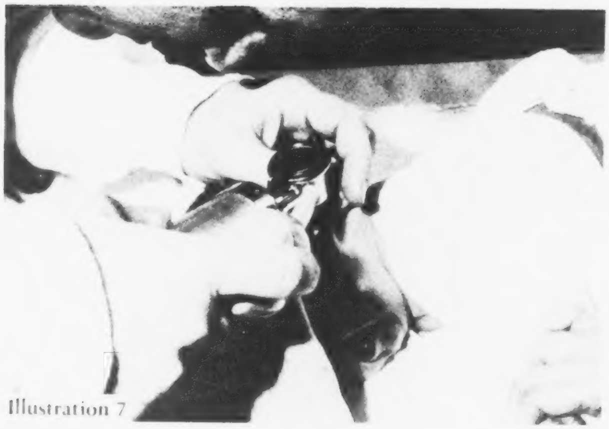

Illustration 7

Illustration 8

Illustration 9

while still taking advantage of two-power magnification. The blunt curette is held lightly and all instrumentation is done slowly using light pressure to avoid scratches of the ear canal. The curette is typically used and cleaned many times on a gauze or cloth while cleaning each ear canal. If a scratch occurs and a tiny drop of blood appears, the examination can still continue but if a lot of bleeding occurs, the examination must be terminated. In either case, three drops of Cortisporin ear drops should be placed in the ear canal at the end of the examination if bleeding occurs to prevent an external otitis from developing.

Pneumatic otoscopy is performed after replacing the otoscope in the right hand, moving the magnifying lens to its closed position, and then compressing the bulb against the handle of the otoscope using the right index finger. *(Illustration* #7)

The examination of the right ear proceeds after the infant's head has been turned to the left and the aide moves to the left side of the table. Now the assistant's right hand is placed on the newborn's forehead to press it firmly against the table while the assistant's left hand holds both hands of the newborn and pulls them away from the head so that the shoulder again cannot interfere with otoscopy. The physician now places the otoscope in the left hand while using the right hand to clear the ear canal. *(Illustration* #8) Pneumatic otoscopy is continued with the otoscope in the right hand while the left hand pulls the pinna inferiorly. *(Illustration* #9)

The neonatal eardrum differs from the adult eardrum and is not just a small adult eardrum. The tympanic membrane of the neonate is as wide as the adult but is not as tall. *(Illustration* #10) This is due to an optical illusion which can be understood by seeing the side view of the ear canal and middle ear as in Illustration #11. The structures seen in and behind the tympanic membrane also differ. *(Illustration* #10) In the adult, the short process, handle, and umbo of the malleus are visible. The long process of the incus and the stapedial tendon may be seen through a translucent tympanic membrane. In the neonate, however, only two landmarks can be seen. The short process of the malleus is a small round nubbin slightly projecting from the plane of the tympanic membrane. The umbo is represented only by a concavity in the center of the tympanic membrane, much like the surface of an erythrocyte. The tympanic membrane of the newborn is gray-pink and opaque. This is in contrast to the tympanic membrane of an older infant or child where the tympanic membrane is silvery gray and translucent.

The landmarks of the tympanic membrane during the examination are upside down because the physician is standing at the head of the table. *(Illustration* #12) In this view, the first landmark to come upon is the slight bulge of the short process because it is anatomically closer to the examiner.

A comparison between the adult and newborn tympanic membrane as seen from a side view reveals that the neonatal tympanic membrane is angulated much more sharply than the adult and assumes more of a horizontal plane, almost in line with the superior portion of the ear canal. *(Illustration* #11) Therefore, although the tympanic membrane is as tall as in the adult, there is an apparent decrease in height due to foreshortening. The differentiation between tympanic membrane and canal wall was initially a problem.

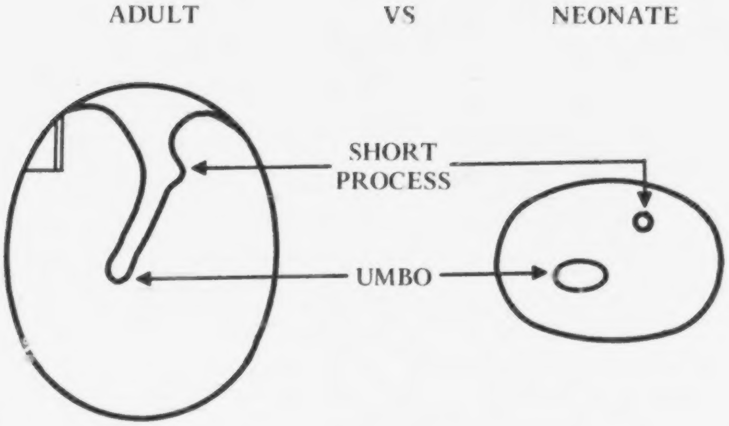

Illustration 10

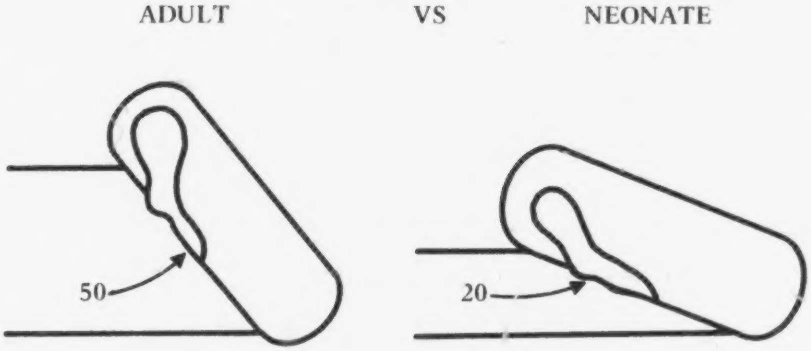

Illustration 11

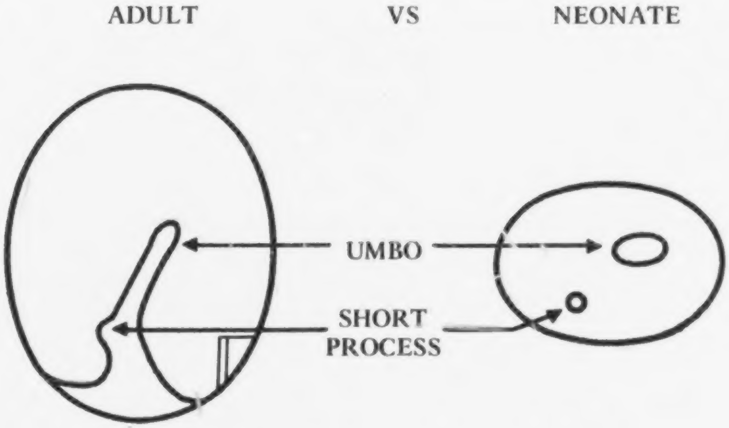

Illustration 12

For example, the tympanic membrane and superior canal wall lie in the same plane, so that it is difficult to tell where the canal ends and the pars flaccida begins. Two features make the separation possible: first, there are vessels visible on the tympanic membrane but not on the ear canal skin; second, with pneumatic compression the tympanic membrane moves, but the superior canal wall does not move. Inferiorly the problem is different. The inferior canal wall bulges loosely over the inferior portion of the tympanic membrane (particularly in the premature neonates), and moves with pneumatic compression appearing like movement of the tympanic membrane; however, inferiorly the canal and tympanic membrane lie at an acute angle, and after a little experience the difference can be clearly detected. Standing at the head of the newborn and aiming the speculum toward the neck helps to "sneak under" the inferior canal skin overhand (although in this position the inferior canal skin is located superiorly).

**II. Mobility of the Tympanic Membrane in the Newborn.** It has been stated that amnionic fluid will fill the middle ear of the newborn and that it will require up to six weeks for the fluid to disappear. Therefore, one would expect that pneumatic compression of the tympanic membrane in the first 72 hours of life should reveal an immobile tympanic membrane. However, after examining 20 newborns within the first 72 hours of life, I found 18 out of 20 to have mobile tympanic membranes. The explanation for good mobility is not clear. Perhaps the amnionic fluid does not reach the middle ear. Or the amnionic fluid does reach the middle ear but with swallowing starting immediately after birth, the Eustachian tube opens and the fluid clears via the Eustachian tube allowing air to pass into the middle ear.

Mobility was graded after examining a great number of newborns. The range of mobility was then graded as 0%, 20%, 40%, 60%, 80% and 100%.

Some newborns had poor mobility of the tympanic membrane. In a prior report, we felt that poor mobility of the tympanic membrane in a newborn prognosticates early otitis media with or without suppurative drainage.[1] Poor mobility of the tympanic membrane may be explained in three general ways. First, there could be *fluid in the middle ear* impeding the inward motion of the tympanic membrane. Amnionic fluid may have entered the middle ear and remained there due to a partly obstructed Eustachian tube or due to viscous fluid. Perhaps serous or suppurative fluid filled the middle ear, resulting from intrauterine ear pathology. No myringotomy was performed in any of these infants, so the type of fluid is unknown. (In four other infants with cleft lip and palate who had immobile tympanic membranes at one to four weeks of age, a myringotomy was performed revealing thick tenacious fluid ("glue ear"); therefore, pathologic fluid can certainly be found in an infant's middle ear at a very early age, and may have been present at or before birth.) Stool and Randall[2] found mucoid material in 47 out of 50 middle ears in 25 cleft palate infants ranging in age from 9 days to 12 months, again revealing the early occurrence of middle ear fluid.

Second, poor mobility may be due to *a thick tympanic membrane*. Normally, the tympanic membrane is thin enough to be mobile. McLellan and Stout[3] have described a thickened tympanic membrane in a newborn with acute otitis media; thus, the thick, immobile tympanic membrane could be a sign of intrauterine otitis media.

Third, *embryonal connective tissue* in the middle ear might impede the motion of the tympanic membrane. McLellan, et al[4], examined histologic sections of the ears from 21 fetuses and 15 infants. The largest aggregate of embryonal connective tissue was in the incudal fossa area. Only in a small fetus (95 mm.) were the ossicles enveloped by embryonal connective tissue, and, as the fetus aged, the connective tissue decreased reciprocally. By the time of delivery, the mesotympanum was free of embryonal connective tissue. It is unlikely, therefore, that the poor mobility was due to embryonal connective tissue filling the middle ear in these newborns.

**III. Otitis Media in the Newborn.** For many years, it has been known from autopsies of newborn infants that otitis media can be present. It was unfortunate that this information didn't stimulate clinicians to diagnose otitis media in the living newborn. Certainly, examining the tympanic membrane of the newborn is relatively difficult because vernix caseosum fills the ear canal, because the external auditory canal is tiny, and because normal landmarks of the newborn tympanic membrane are different and not widely discussed. However, each of these problems can be overcome and the disease may become obvious.

There are two general comments about newborn otitis media that have been true in my experience. First, the otitis media is typically bilateral. This bilaterality must be respected for it signifies a definite hearing loss for the newborn infant with probably slight to moderate hearing loss.

Second, the otitis media tends to persist for months and perhaps years, rather than resolve spontaneously. Because the first 18 months of life are so critical for the perception of sound and the development of speech and language, we could predict that this high risk group of infants will probably develop poor language and/or speech.

The diagnosis of otitis media hinges on the fact that the middle ear is filled with an exudate, thereby reducing tympanic membrane mobility. The tympanic membrane may also be pink-red and thick.

I personally examined 101 newborn Navajo Indians[1] and found a mobile and gray-pink tympanic membrane in 71. None of these newborns with a normal ear examination had suppurative otitis media before five months of age and only four of these infants (6 percent) developed ear drainage from 5-7 months of age. However, in the 18 infants with a poorly mobile tympanic membrane, four of them (22 percent) had suppurative otitis media before five months of age. The risk of serious middle ear infections is therefore four times as great in an infant with an immobile tympanic membrane at birth. It is assumed that the immobile tympanic membrane is due to fluid in the middle ear as seen with otitis media. Also in the group with poorly mobile tympanic membranes the risk of early acute otitis media is twice as common as in the group with an initially mobile tympanic membrane.

There have been three high risk groups associated with newborn otitis media:

*1. The Newborn with a Cleft Palate.* Approximately 95 percent of all cleft palate newborns have bilateral otitis media. This can be detected clinically by pneumatic otoscopy and proven by myringotomy. At operation the tympanic membrane is found to be very thick, the middle ear is filled with viscid "glue" or mucoid material, and the mucosa of the medial wall of the middle ear is thickened and polypoid. At one year of age, approximately 90 percent of cleft palate infants have bilateral otitis media; at two years of age approximately 75 percent of cleft palate infants have persistent otitis media. Thus, a prolonged middle ear infection can be anticipated in most cleft palate newborns.

2. *The Premature Infant.* In my survey, premature infants (under 2500 grams) frequently have poorly mobile tympanic membranes. In the Navajo newborns, early suppurative otitis media occurred in 17 percent of the premature infants but in only 8 percent of the full term infants. Prematurity alerts the physicians to a great number of diseases, and perhaps middle ear pathology should now be added to this list.

3. *Amnionitis.* McLellan et al[5] showed an association of amnionitis and neonatal otitis media. It could be anticipated that since amnionic fluid is normally swallowed by the fetus, some of the infected fluid could be pumped into the middle ear during swallowing, thereby establishing an intrauterine otitis media. Antibiotic therapy in the newborn apparently controls this type of otitis media[1].

**IV. Structural Anomalies of the Ear Canal and Middle Ear.** Embryologic development of the ear canal and middle ear is a complex process terminating in a thin, delicate tympanic membrane and three ossicles, perfectly joined together, with a demand for minimal margins of error in development in order to transmit sound waves via a mobile coordinated unit of tympanic membrane and ossicles into the inner ear. In general the factors affecting abnormal embryogenesis have not been identified. Only the following few specific hereditary or congenital categories have been identified where abnormal embryogenesis occurs in the conductive system of the ear:

1. Hereditary familial patterns have only occasionally been identified. For example, footplate fixation has been found in three families with an X-linked dominant transmission[6]. An additional family was identified with middle ear ossicles fixed by bony bridges and a G-group chromosome depletion was identified[7]:

2. Thalidomide has been associated with congenital conductive losses by producing footplate fixation or canal atresia[8].

3. Rubella is an example of a viral infection that may lead to the first arch syndrome with canal atresia, or to congenital fixation of the stapes footplate[8].

### Classification

A descriptive compilation of the possible structural anomalies with brief descriptions is included to show the very wide spectrum of pathology that exists. Four groups have been categorized as follows:

A. Isolated Middle Ear Anomalies

B. Isolated Aural Atresia

C. Middle Ear Anomalies with Associated Congenital Anomalies

D. Aural Atresia with Associated Congenital Anomalies

Groups A and C newborns are difficult to identify at birth since the ear canal is normal and the tympanic membrane may be normal. Group A newborns (Isolated Middle Ear Anomalies) are the hardest to identify since there are no external clues that an anomaly of the middle ear is present. It is unlikely that these anomalies will be identified in the newborn except in a most unusual situation. Group C newborns (Middle Ear Anomalies with Associated Congenital Anomalies) is a rapidly expanding group. At the present time it would be appropriate to consider a middle ear anomaly as possibly occurring with any other congenital anomaly.

Groups B and D newborns are easy to identify since the external auditory canal is absent at birth. In these cases, auditory rehabilitation is the key to management.

A. **Isolated Middle Ear Anomalies.** Conceivably the ossicles could be individually or collectively deformed, deficient or fused to the surrounding bony walls – each producing a conductive hearing loss. Some of the more common examples are discussed below.

*Malleus. Fixation of the head of the malleus* is the most common anomaly of the malleus. The point of fixation may be medial, lateral, anterior or superior to the malleus head. The mechanisms causing the fixation have been described as excessive air cell formation in the epitympanum[9], a long bony septum in the epitympanum[10], partial failure of the epitympanic expansion[10], and bony fixation of the anterior malleal ligament[11].

The clinical diagnosis in the newborn will probably never be made. However, in the infant or child the diagnosis is established by using the pneumatic otoscope and noticing that the malleus is immobile even though the tympanic membrane is mobile.

Palpation of the malleus at operation will produce no motion, although the incus and stapes are mobile. Surgical repair by 1) enlarging the attic space or 2) dividing the malleus head and performing an incus interposition are sound approaches.

*Incus. A deficiency of the incus* is the most common anomaly of the incus. The deficiency may range from an absent lenticular process to total absence of the incus[12]. An appropriate ossiculoplasty for each anomaly must be entertained, and may range from a bone chip wedge between the incus and stapes, to a total incus replacement by a bone graft or prothesis.

*Fixation of the incus* in the attic may also occur[9]. If the fixation occurs at the tip of the short process merely dividing the short process will mobilize the incus. If the body of the incus is fixed, enlarging the attic space is necessary.

*Stapes. Footplate fixation to the otic capsule* is a common form of congenital conductive hearing loss. Embryologically the footplate is fixed but must undergo differentiation of the mesenchyme at the future annular ligament to allow for motion. A failure of differentiation is the cause of the fixation.

At operation the stapes may be mobilized if the fixation is fragile, and in more rigid fixations a stapedectomy is needed, without or with a drill-out technique.

*Oval Window. Absence of the oval window* may be considered a more severe form of footplate fixation, and it is associated with anomalies of the stapes. Reconstruction is difficult due to injuries of the facial nerve, difficulty finding the vestibule, perilymph "gushers" from the vestibule, and injury to the inner ear.

**B. Isolated Aural Atresia.** An absent ear canal is usually associated with anomalies of the pinna and the middle ear since all three are derived from the first and second branchial arches. The pinna deformity may range from mild cartilage deformity to a small pinna (microtia) or to an absent pinna. The ear canal is usually totally atretic, but in some cases the canal is atretic only in the medial half. A stenotic ear canal should be considered a minor form of aural atresia since both conditions may occur in the same infant, and because both stenosis and atresia are associated with similar pinna and middle ear deformities.

The middle ear anomalies are quite varied. The malleus and incus are often fused together, and appear as a bulky stubby mass of bone. The fused ossicles may be mobile or fixed to the middle ear. The stapes superstructure is often deformed.

The etiology is usually unknown. There appears to be a low hereditary pattern. It is very uncommon to see aural atresia in families or siblings. In my Navajo Indian experience only two siblings were involved in 56 patients and no parent-child pairs were encountered with aural atresia. This low heredity pattern is consistent with the experience of others. Bilateral atresia was present in two of 56 patients, and appeared to be lower than the 10-20 percent prevalence usually reported. Thalidomide and rubella infections in pregnancy have also been associated with aural atresia.

A hearing aid may provide the amplification during the speech and language formative years until the atresia is corrected in early childhood.

Operative correction of atresia is one of the most difficult otologic challenges. Therefore operative correction should be contemplated in cases with bilateral atresia or selected unilateral cases. The problems encountered in surgery are (1) how to avoid injury to the facial nerve, (2) how to reconstruct the ossicular chain, (3) how to prevent canal stenosis and (4) is it advisable to reoperate in the patients who fail to regain a satisfactory hearing level? The otologist's skill and experience will dictate the answers to these questions, since firm doctrines are not available.

**C. Middle Ear Anomalies Associated with Other Anomalies.** Many anomalies of the middle ear are associated with other anomalies or syndromes. At the present time, I believe that any newborn with any anomaly should be considered as possibly having an associated congenital hearing loss. Until evidence is accumulated to exonerate certain classes of anomalies from being associated with hearing losses, I would recommend that all these newborns be seen by an otologist. Serial examinations and hearing tests will probably be needed to confirm or deny the coexistence of a hearing loss and when needed, appropriate rehabilitation could be instituted as early as possible.

The following list has been compiled from the literature and my personal experience and represents those congenital anomalies which have been found to have an associated middle ear anomaly:

1. *Pinna*

    low set
    thickened
    cupped and preauricular pits
    cupped and preauricular pits and cervical fistulae

2. *Face*

    hemifacial atrophy (1st arch syndrome)
    mandibulo-facial dysostosis (Treacher-Collins syndrome)
    craniofacial dysplasia (Pyle's disease)
    otofacial cervical syndrome

3. *Skeletal*

    osteogenesis imperfecta
    osteopetrosis
    symphalangism
    cleft hands
    Turner's syndrome
    Klippel Feil syndrome
    Spengle's deformity
    Madelung's deformity

4. *Chromosomal anomalies*

    G-group depletion
    XXXX

5. *Cleft palate*

    otopalatal digital syndrome

6. *Eye*

    Moebius syndrome

7. *Skin*

   knuckle pads and leukonychia

8. *Endocrine*

   cretinism

**D. Aural Atresia Associated with Other Anomalies.** The newborn with a congenital anomaly and atresia of one ear is obvious to identify. However, it is important to point out that the "uninvolved" ear with a normal ear canal may also have an anomaly of the middle ear. As the infant grows and mental retardation becomes obvious, the retardation may be attributed to a CNS anomaly rather than to the bilateral hearing loss that actually exists. Therefore, any newborn with a unilateral atresia should have an otologic and audiologic evaluation to assess the hearing status of both ears.

The following list has been compiled from the literature and my personal experience and represents those congenital anomalies which have been found to have an associated aural atresia:

1. *Face*

   1st arch syndrome
   1st and 2nd arch syndromes
   mandibulo-facial dysostosis

2. *Skull*

   Crouzon's syndrome
   Apert's syndrome

3. *Eye*

   Goldenhar's syndrome
   Duane's syndrome
   Moebius' syndrome
   congenital bilateral abducens nerve paralysis

4. *Skeletal syndrome*

   osteopathia stria and osteopetrosis
   Klippel Feil syndrome
   cleidocranial dysostosis
   acromegaly (cystathioninuria)

5. *Cleft palate*

6. *Renal-genital*

7. *Other*

Wildervanck
Thalidomide

## SUMMARY

The newborn infant can be born with a conductive hearing loss. When bilateral losses occur, speech and language may be impaired.

The technique for examining the tympanic membrane in the newborn is discussed in detail.

The use of the pneumatic otoscope is stressed so that mobility of the tympanic membrane can be determined. At the present time, it is believed that poor mobility of the tympanic membrane in the newborn represents an abnormal condition and it is a prognostication of early otitis media.

Otitis media can occur in the newborn. Three high risk categories where otitis media is found are (1) newborns with a cleft palate, (2) premature infants and (3) delivery associated with amnionitis.

A diversified group of structural anomalies giving a congenital conductive hearing loss can occur. In the obvious cases, only the middle ear structures may be deformed. It seems appropriate to consider a middle ear anomaly when any congenital anomaly occurs.

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# THEORETICAL CONSIDERATIONS IN THE SELECTION OF VARIABLES FOR TESTING THE HEARING OF NEWBORNS

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Our purpose is to develop and explore a theoretical or conceptual framework that will identify the many choices which an experimenter must make when he measures "hearing" in newborns. We would like to introduce the word "transaction" to help our understanding of the complexities that exist between an observer and a subject during any test of hearing. The word "transaction" will be used because it emphasizes the multiplicity of interactions which are in progress and because it brings out the concept that the whole is probably more than the sum of the interactions that it comprises. Regardless of how much one wishes to remain alert to the dynamics of the entire situation, it is all too easy to drop into the consideration of how some small subset of variables are operating. This is presumably related to the limitations of the human mind in processing many variables simultaneously. For this reason it is worthwhile to attempt to envision the whole of the auditory test situation many, many times. From each review we can gain clarity. This clarity is particularly useful in the ever-changing problem of testing newborns. For newborns bring to the situation variables which are fully as complex as those in adults and at the same time less capable of being set or recognized at any one given moment.

One can best look at the newborn auditory test situation by imagining that one is viewing it from somewhere in an N dimensional space-time continuum. Looking from here, we would see two spheres of influence, one the subject, the other the experimenter. Figure 1 shows two circles representing these spheres. Each sphere is in dynamic exchange with its environment. As the test situation develops the two spheres become confluent in an area which forms the interface across which their transaction occurs. In Figure 1 the interface is the area of overlap of the two circles. The transaction continually modifies the feedback of both experimenter and subject. Even though a broad range of exchanges can and does transact across the interface we see the experimenter concentrate his effort on a selected subset of his outputs and inputs. In Figure 1 the interface is broken down into four areas and the list under each gives representative kinds of variables. The set the experimenter chooses acts like a filter of experience and modifies the emphasis and scope of the whole transaction.

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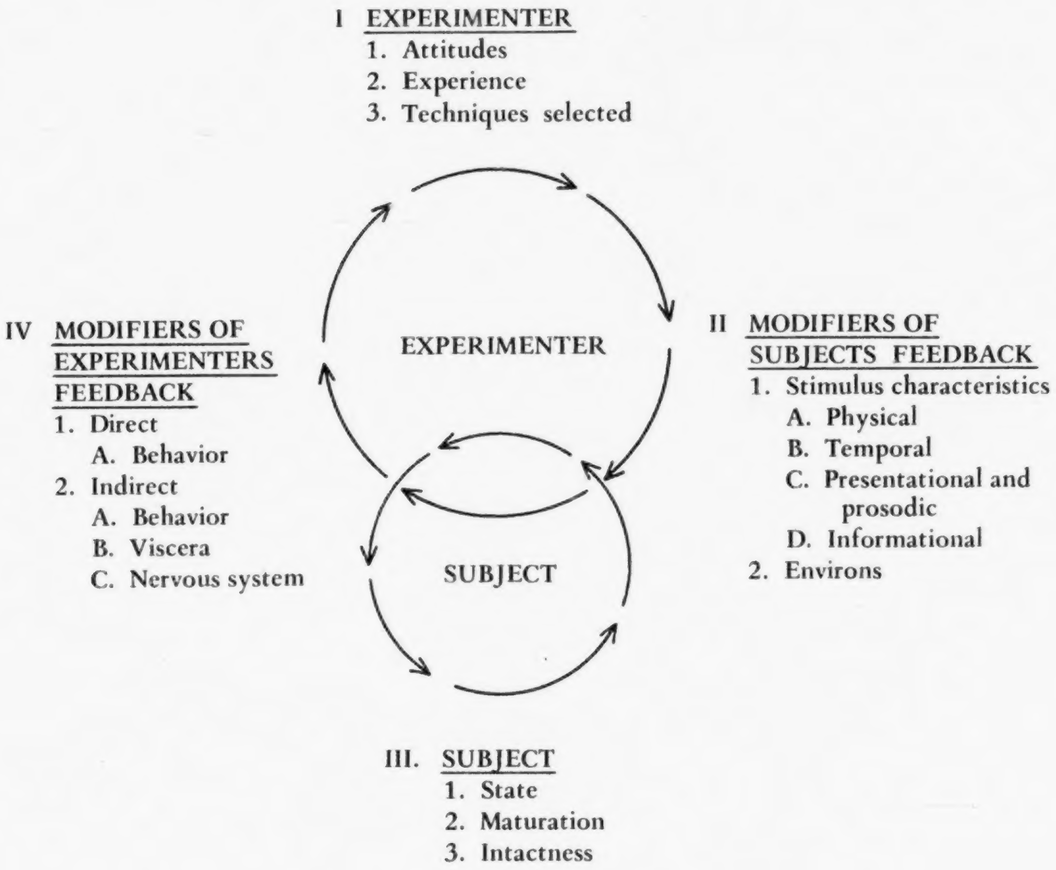
Figure 1

THE EXPERIMENTER-SUBJECT TRANSACTION THAT DEFINES "HEARING"



The experimenter cannot ask the newborn to concentrate his efforts on some set of variables. In fact it is often not possible to get cooperation in this sense even from adults without training and motivation. The newborn therefore imposes his biases as well upon the transaction. The infant's motivation and goals often vary rapidly. If he is interested in food and receives only sound, the infant may define himself in a way which greatly limits his range of responses often turning his set from the situation to his need for food and back and forth unpredictably. If we allow that the type and range of responses can vary depending upon the infant's self-definition by his goals, drives and motives, then the experimenter must be prepared to transact on widely differing bases or else to accept widely varying success with a single set of criteria of response.

Dr. Eisenberg[1] has documented the wide range and the importance of such factors in hearing testing of infants using behavioral criteria for response. Dr. Rapin[2] has documented the same using the electrical output of the brain as evidence of response. It matters little what area we look at, the transaction is modified by the set of the infant. Even the cochlea receives a significant motor innervation via Rasmussen's bundle and if this operates to modify the neural firing from the cochlea as Galambos[3] has suggested, then even the periphery need not escape the effects of the set of the total transaction.

But just as the set of the infant modifies the transaction, so does the set of the experimenter. To begin with the experimenter has a very wide range of variables from which to select the ones he will accept as measuring hearing. The ear is the sense organ with the widest connections within the central nervous system. From the cochlear nuclei at the side of the medulla there are connections to all of the varieties of subsystems within the brain. To paraphrase lectures of Stanley Cobb hearing stands guard while the animal eats or sleeps or performs other self-oriented acts. The system is capable of resetting the entire central nervous system to adjust to acoustic and therefore distant signs of danger. Sound is capable of producing changes in any or all portions of the organism. There can be changes in heart rate, peripheral circulation, sweating, respiration, in movement of eyes, face, limbs or entire body, complex acts such as sucking, crying, babbling, may increase or decrease and if one accepts consciousness, sounds reach and modify its states.

Because of this breadth of possible responses the experimenter usually selects some particular cluster as his indicator. He can't reasonably be expected to look everywhere at once. In doing this he increases his confidence in his results but, at the same time, he defines what he will accept as "hearing."

This is why we have put the word "hearing" in quotes and why we will continue to do so. There is a different neural path for every different response. Each response can therefore be interfered with by a different disturbance. One would imagine that the cochlea would be in common for all these different responses but the cochlea is not that simple. It is highly differentiated into inner and outer hair cells; into a range of structures from base to apex; it is probably locally modified by the efferent paths to it, so that even here there may be differential action.

On this basis what each experimenter accepts as response does define what "hearing" he is measuring. Since no one can measure all the variables then no one measures the total hearing. One can only measure some hearing. Several conclusions result from this argument and they are worth exploring. The essence of this discourse is that two questions should be kept uppermost in our minds. One is, "What hearing does any given test measure?" and the other is "Is that the hearing that I wish to know about the newborn in question?" This problem was described in 1958 by Harris.[4]

If we had reports from several different observers we would no longer expect them to agree. If each used a different criterion then the hearing measured should be different. Each form of test therefore needs to have its own standard of reference.

This raises the old problem of validity and reliability for a test. Validity is evidence supporting the fact that the test measures what it is trying to measure. Reliability is an estimate of the probability of obtaining the same result on a repeat test or on another subject with an identical problem.

For example, it is important to ask is this test a valid measure of cochlear function or does it test the connection of the auditory paths to the reticular system or does it involve both of these functions? Validity for each test would be established on a different experimental basis.

One important outcome from consideration of validity is that it highlights an error we make when we test out a new procedure by comparing its results with an older, well-established procedure which does not test the same "hearing."

For example, one often finds attempts to prove the validity of EEG evoked responses by comparing its results with those of standard audiometry. The only time these two must necessarily agree is when that part of the auditory system which they share in common is the only part disturbed or is the only limiting factor of the function. Since we can't define precisely how much of the system is in common other than peripheral ear, and cochlea, the comparison is not useful.

When we deal with patients with disturbances exclusively in the peripheral ear the comparison could be helpful but even here there is room for doubt. The absence of peripheral cochlear input to the brain could have resulted in a totally different responsiveness between the evoked potential system and the voluntary response. The two systems could adapt differently to this "deafness." There is evidence of this in that deaf preschool children who have had no auditory training have lower EEG thresholds than audiometric thresholds. This reverses after a year of auditory school experience. If the two tests to yield exactly the same results in all cases then probably something is very suspicious about our basic concept of either or both standard audiometry and EEG evoked responses. One or both must be operating on a set of unrecognized factors.

The reliability of a test develops the concept of the probabilistic world in which we live. It develops our estimators of how much signal is buried in how much extraneous unwanted activity or noise. We think that it is necessary to express findings about "hearing" in these probabilistic terms. In our communication across disciplines it is essential to be informed as to the chances of error – the false negatives, the false positive as well as the confidence one can place in any finding or interpretation. Much of miscommunication would be avoided if we were all careful to couch our reports in terms of probability.

In this argument we have skirted frequently on the edge of the concept of cause and effect. We turn on a tone and it causes the organism to respond. The simplest form of this idea is found in the reflex arc. If a certain energy is applied a predictable response will appear. While this has been and can still be a useful concept it has several problems inherent in it. One is that if this defines the subject, doesn't it also define the experimenter? The subject moves so the experimenter presents another stimulus. And now the experimenter is the responder because he reacted to the subject. Logic of cause and effect such as this leads to curious results such as the idea that sensation is passive. Eventually, that all the elements we view from our N dimensional setting are passively linked together like a chain of dominoes standing on end.

We object to being considered as passive experimenters or passive subjects. We believe that there are active processes within every living organism which listen and which develop action. Years ago G. Bishop[5] described a neural circuit that could be a physiological model for such a system. He found that nerve impulses in the visual tracts of the rabbit would not pass through the lateral geniculate body to the cortex unless they met at this body by a synchronized facilitating or unlocking neural discharge from the cortex. It is as if the rabbit would be cortically blind if he did not look. Brunner, Postman and many others have also promulgated this argument. Sensation is probably not a passive reaction. The same is undoubtedly true of the motor output or behavior. Consequently in an auditory test situation what we search for is a modification of our feedbacks from the transaction with the subject that tell us that on the basis of the past ongoing activity a nonpredictable event has occurred in association with our output. What this means is that an observer develops a kind of Gaussian probability for what is likely to happen and it is when this expectancy is violated that one recognizes that a response is occurring. If the subject is listening and formulating response patterns as an innate part of his being then the experimenter's problem becomes more complex if he is to detect which is reaction and which is only the likely ongoing activity. This approach may seem complicated but we think it is essential because a full understanding of the situation comes only through being able to state the proposition in general.

When we referred above to the concentration of the experimenter on certain of his feedbacks we had in mind the possibility of his concentration on many components as well as on total behavior. We meant to include such procedures as recording from electrodes in muscles, on scalp, over sweat glands on arms and legs, etc. We meant also to include the use of special oscillators, tape recorders and other electronic devices by which the experimenter modifies the interface of the transaction. With the use of these instruments the case for selection of what is hearing by the experimenter is most obvious. It has been common to refer to the use of these instrumental procedures as "objective" tests. We wish to take strong exception to this word "objective" and to substitute the word "indirect." A direct transaction means a testing of some hearing functions by confrontation of experimenter and subject. It implies the use of man-made sounds as the experimenter's output and the use of direct sensory detection by the experimenter of the subject's related behavioral changes. To interpose instrumentation between subject and experimenter transferring energy in either direction is not to make the process less subjective only to make it indirect. Because of the innate characteristics of man there are limitations of his outputs and inputs. Instrumentation allows him to exceed these limits. It also allows him to become much more precise and define much more critically either an input or an output. So instrumentation has both advantages and disadvantages. It nearly always makes the "hearing" measured far more limited to one narrow aspect of it. Instruments allow one to know more and more about less and less.

Instrumentation does not make the process less subjective. To begin with, the experimenter has chosen the instruments. He also set up the criteria to be used in determining whether he received signal or noise. The experimenter analyzed the data. The experimenter is the one who writes the report in words which contain connotative as well as the intended denotative meanings. All this is a very subjective process.

There was at one time a whole school of thought based on the concept that the subject's verbal reports were not objective data and that only objective data were good. Yet all the reports of this school were verbal and subjective. What we must recognize is that direct and indirect forms can exist within the total possible transaction between subject and experimenter. We must use no pejorative terms such as subjective and objective. We must look at what we are really doing without fear and with the hope that through understanding of the process we will become more aware of the fraction of the total that we are calling hearing. With indirect measures the complexity and sophistication increases and the results become more specific. It also becomes more difficult to interpret what that particular measure means in terms of the total hearing act.

Before we leave our N dimensional space let us look at how we could define hearing from this vantage point. We would ask ourselves what components of the totality of the transaction that we are observing we would accept as "hearing." Assuming we know what sound is we would accept as hearing those parts of the transaction which would not occur if sound were absent. (1) We could evaluate the transaction by the changes due to sound which occur in the subject, just as the experimenter does. (2) We could just as well follow the changes in the experimenter's output. If he uses a completely inflexible pattern of presentation of sound to the subject we can only conclude that the experimenter is behaving as if only one very particular item were hearing. (3) We could also observe the subject over some time interval and see if he had learned something; that is, was different in his maturity, learning or adaptation because of the auditory aspect of the transaction. (4) We could also observe the experimenter and see if his maturity, learning and adaptation were modified by his experience. Are not all of these in one or another sense perfectly respectable measures of hearing? We have all experienced leaving a test situation with a feeling about the subject just tested. It is difficult to shake such a conviction if the data analyzed later should disagree with that impression. It would be easier from our N dimensional view to separate out how much of this is experimenter determined and how much is transaction determined.

There is a form of statistics which is not only generally useful but also highlights for the experimenter the effects of his choices and attitudes. It is called sequential analysis[6]. In this statistic the experimenter begins by specifying four values.

1. $P_1$ = The maximal percent of failure allowable in any sample of test trials and still consider the sample as acceptable in this case as evidence of responsiveness to sound. Let us assume a newborn hearing test situation. The question asks "What percentage of the time would you allow the infant to fail to respond and still consider him to "hear" the sound? We propose 15 percent. That is, in 20 trials we would allow three failures and still consider that he responds.

2. $P_2$ = The minimal percentage of failure that we would allow and still be confident that there was no response. Obviously if he responded to none of the 20 we would

say that he didn't respond. How about one response in 20 (5 percent)? Is that still no response? How about two in 20 (10 percent): As you proceed with this argument you learn a great deal about the experimenter. If he sets $P_2$ as 5 percent (1 positive) then the differential between 5 percent positives and 85 percent positives ($P_1$) becomes relatively easy. In just a few trials this differential can be determined but is it realistic? If there is lots of unelicited behavior from which one must detect response then the system can easily include nonresponders with the responders. If experimenter says 16 percent failures is not hearing then the distinction between responders (15 percent failures) and nonresponders (10 percent failures) may take an extremely high number of trials. So one either determines the percentage of positive responses in known "deaf" infants or one makes a logical guess. Let us guess that a 50/50 chance of response to any one stimulus is not unlikely in any infant and that the maximal failure to respond that we would allow would therefore be about 60 percent. We are then saying that a child responding 85 percent of the time "hears" and that a child responding 40 percent of the time doesn't "hear."

3. $\alpha$ = Now the experimenter must determine the alpha error he will live with. This is the risk he will run of rejecting his hypothesis when in fact it is true. On Columbus' voyage, since in truth there was land ahead he would have committed an alpha error if he had turned back. Let's accept 5 percent for this value. It is not uncommonly done.

4. $\beta$ = Now the experimenter selects the beta error he will live with. This is the chance he will take of accepting a falsehood. In Columbus' voyage this would have been to sail on when the truth was that there was no land ahead. This value is usually not measured in most statistics and rarely set by the experimenter. Let us accept 10 percent for this value.

What sequential analysis does is to operate on the sample as it grows trial by trial. It determines after each new trial which of the three possible categories was achieved within confines of alpha and beta errors.

1. The sample meets the requirements of $P_1$.

2. The sample meets the requirements of $P_2$.

3. The sample lies between the two (in our case between 60 percent and 15 percent) and one should continue sampling.

In this statistic N becomes a variable and collection proceeds until a solution is reached. There is the possibility that one might have to continue sampling for an unreasonable number of trials because of the choice of the four values. There is a calculation to determine if this is true and even better there is a calculation of the average sample number once given the four determinants.

Figure 2

$P_1 = 15\%$ $P_2 = 60\%$ $\alpha = .05$ $\beta = .10$

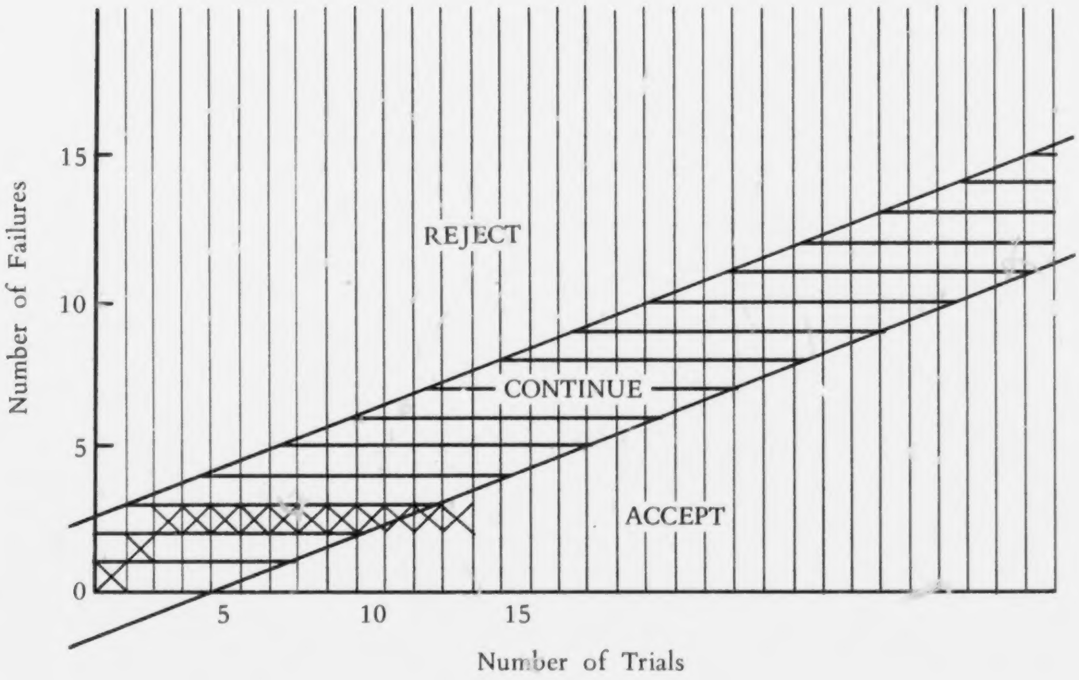


Figure 2 is the graphic solution to our problem. The slope of the lines and their distance apart (the intercepts) were calculated from our four values and plotted on coordinate paper. On the graph are also represented the result of 13 consecutive observations (X). The results can be plotted as the test proceeds. Every trial is plotted to the right on the abcissa of the previous one. It is also plotted one space higher on the ordinate scale whenever it is a failure. In Figure 2 the first three trials were failures (the first test is always plotted in the same place whether a success or a failure). The remainder were all successes until at the 12th trial when the entries crossed into the zone of acceptance of the sample as satisfying the criteria of "a responder." At this point sampling can cease. This procedure is efficient. It allows one to center his effort in those situations where the distinction is difficult. The setting of the four determinants describes much about the experimenter. The data obtained describes the way in which the subject relates to the experimenter's criteria.

The transaction we have been discussing is floating within a still greater field of influence, namely society. There are pressures which modify experimenter, subject and all the people and equipment related to this transaction.

Society, for example, may set up legal definitions of what constitutes a test of hearing. The experimenter may find himself practicing defensively and performing tests which he doesn't consider necessary merely to satisfy the legal requirements. Society, even the experimenter's scientific society, may be going through a phase of being excited by a particular approach. These pressures will modify the selections that the experimenter makes. Society also puts pressures on the newborn to sort of "shape up" and hear like the rest of us. Life is not geared to accept with equality all types of deviations from the mean. The child's mother may have strong pressures brought to bear on her through her knowledge about deafness. Her knowledge may be limited, distorted, or even archaic. It is often less likely to be mature and modern. Therefore she has modified the infant whenever she brings anxiety to certain types of relationships with him. When the mother meets the experimenter she often transfers these pressures to him or at least places them before him and he may respond if he is a passive receiver.

Society also places great pressures on this situation by its response to the determination of deafness in the newborn. Only some localities have adequate training situations. There are communities whose facilities are so good that parents of deaf children move into this locality in order to obtain this educational opportunity for their child.

Society also puts a cost on all things. So for the interest of it we have made a rough cost accounting of the identification of a deaf newborn. Perhaps our figures are not right but the formula is accurate.

Let us assume that if a child were identified as deaf in the first six months he would have a better language development because of the use of hearing aids and training during the critical ages of one to three. Let us further assume that as a result he could earn $1,000 a year more as an adult than if he had not been detected until, say, four years of age. Allowing a 30 year period of adult productivity, the early identification of deafness developed a $30,000 increase in earning capacity of the subject. Let us train someone to make these identifications in infants. They would have to identify one such infant every year in order to create this $30,000 fund. Let us pay the tester $20,000 per year and use the $10,000 for equipment, supplies and overhead of depreciation of equipment. Since the problem occurs once in every 5,000 births, such a tester would have to do 5,000 studies a year plus, probably, 200 repeat studies on positives identified. If there were 240 work days available, this would mean doing almost 22 studies per day (5,200/240). On the basis of a six-hour day, this is about 17 minutes allowed per test. We would, by the way, reduce the fund created every time a false positive was processed because some parent went through needless anxiety (child called deaf and wasn't).

We have covered a wide range of factors which contribute to the measurement of hearing in newborns. Developing the very awareness of the width of these variables was our goal on the one hand while on the other we hoped to offer ideas for handling them. Since one cannot hope at this point to measure all of hearing, the path before us is to select for measurement one aspect that can be helpful in either broadening the life of our subject or broadening knowledge about such subjects.

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# CURRENT OVERVIEW OF NEWBORN HEARING SCREENING

Marion P. Downs*

It is no coincidence that the papers which have been presented at this conference deal largely with the High Risk factors which accompany deafness. Although Dr. Konigsmark has estimated that 30 to 40 percent of all congenital deafnesses may not be able to be identified on a High Risk Register, the figures that I will present indicate a probably much lower figure in actual practice. Indeed, I will attempt to demonstrate in this paper that the most feasible approach to newborn hearing screening at the present time may be to do careful hearing screening tests on only those newborns who fall into a High Risk Register.

However, before that I would like to comment on the silent assumptions that have underlaid the papers and discussions at this conference: First, the assumption that it is of the utmost urgency to identify hearing loss at as early an age as possible; second, that critical periods do exist for the development of language and of hearing perceptions, and that devastating effects result if the child is not given auditory stimulation or language input at the proper time; and third, that the expertise exists to test the hearing of the infant at birth, with some form of diligent professional observations and instrumentation. These are all valid assumptions, and indicate that we have come a long way in our thinking in the last twenty years.

What remains is to determine the most effective procedure for accomplishing the identification of deafness at birth. Many statistics have been published on the numbers of deaf infants found at birth through mass screening programs, using observations of behavioral responses.[1] These statistics show that various programs have detected everywhere from one deaf infant in 1,000 to none in 14,000; out of 61,000, an incidence of 1:2800 was reported; and that three false negatives (those who passed the screening test but who ultimately appeared with deafness) were reported in this number. A 12 percent average of false positives were found (those who failed the screening but who were normal hearing).

But these statistics hide some vital facts: when one looks at the etiologies and pathologies of the hearing losses in the programs which list these factors, it appears that most of the infants identified would have been, or were, on a High Risk Register. Furthermore, when one looks carefully at the false negatives reported, it appears that some among them may have developed deafness well after birth. To illustrate these facts, I would like to present the results of our own hospital's testing since 1964, of approximately 20,000 newborns, and then to refer to Dr. Feinmesser's study from Israel. The children I report are known intimately to us – some of them for as long as six years, and we are certain about their degree of deafness and the etiologies of their losses. The infants who were identified by screening are listed by the following etiologies:

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|  |  |  |
|---|---|---|
| 1. | Recessive profound deafness with one known deaf sibling | High Risk |
| 2. | Maternal rubella severe deafness | High Risk |
| 3. | Recessive severe deafness with two known deaf siblings, one with renal syndrome | High Risk |
| 4. | Possible recessive profound deafness with no known family history, but Rh negative mother | High Risk |
| 5. | Apert's syndrome, conductive loss | High Risk |
| 6. | Microcephaly and microtia | High Risk |

These are the surviving children identified by screening at birth. There is another group of infants who were identified as deaf and who expired within short periods after birth. On all but one the temporal bones were obtained and the histopathologic studies have been or are being reported in the medical journals. These are:

|  |  |  |
|---|---|---|
| 7. | Osteogenesis imperfecta[2] | High Risk |
| 8. | Partial deletion of chromosome 13[3] | High Risk |
| 9. | Trisomy 13-15[4] | High Risk |
| 10. | Maternal Rubella[5] | High Risk |
| 11. | Treacher-Collins syndrome[6] | High Risk |
| 12. | Jervell-Lange Nielsen syndrome | High Risk |

Every one of these infants would have been on a high risk register had one been in effect during this entire period. It is evident that the screening tests performed on the non-High Risk group were not very productive.

The false negatives in our program – those who were cleared at birth but who later appeared with deafness – give us equally vital information. These consist of:

1. Recessive deafness with no known family history of deafness: tested at birth and again at three months of age and cleared of both tests. At eight months the patient presented with profound sensori-neural deafness. It is considered highly probable that the deafness developed between three and eight months of age.

2. Recessive deafness: cleared at birth, but presented at six months of age with profound deafness. In the absence of an intervening test this case must be considered as a failure of of the screening procedure.

3. Recessive deafness with known deaf sibling: cleared at birth but at one year of age appeared with severe sensori-neural deafness.

Although these three may all represent screening failures, at least one of them probably had normal hearing until three months. We cannot overlook the possibility that these recessive deafnesses may well have developed the losses at some time after birth. We have one documented case of the development of such a hearing loss.

Figure 1 shows the progressive audiogram of a child with two older deaf siblings, one normal sibling. This child was tested regularly during infancy and was deemed normal. At two years of age he was able to give us good play-conditioned responses, and the top contours show his thresholds at that age. (Bone conduction was identical with air conduction at all tests.) Two and three years later there was progression in all frequencies, but still essentially good hearing. Normal speech and language development obtained at that point. Two months later the hearing had fallen to a severe deafness, and within another two months the loss was profound. This case gives an interesting documentation of how and in what period of time a recessive deafness can develop.

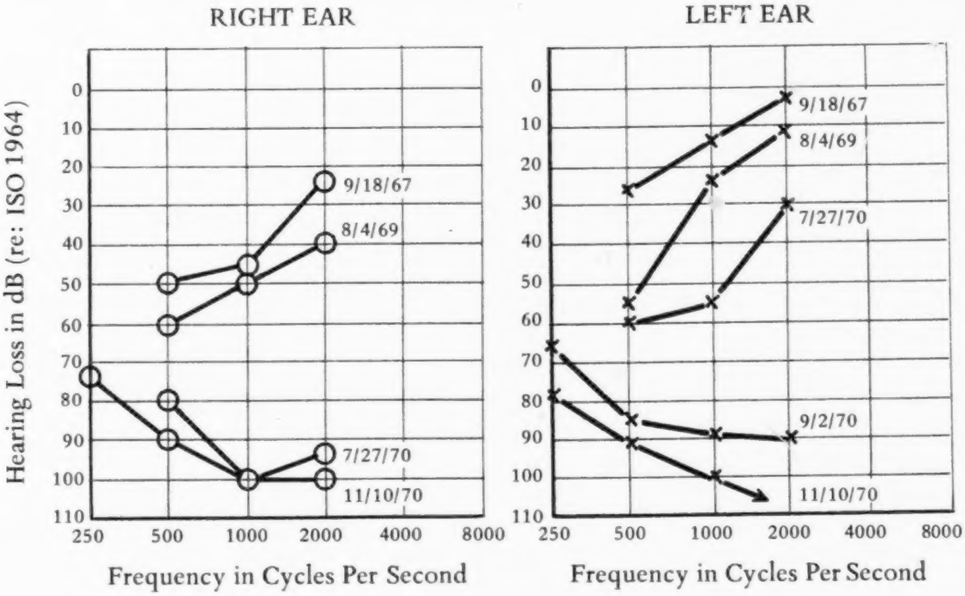
The most comprehensive screening program that has been reported is detailed in Dr. Feinmesser's paper which appears as Appendix A. This project at Hadassah Hospital in Jerusalem has screened 17,000 newborns to date, but his present report is on 10,000 infants. Not only were these infants screened by observations of auditory behavior, but 20 percent of them were placed on an inclusive High Risk Register. More than 80 percent of the entire 10,000 were retested at well-baby clinics at seven months and again at 18 months. These babies were also given developmental tests. On the last page of his paper are shown the results: Of nine deaf infants in the population, only one was not on the High Risk Register. The behavioral screening identified five of the deaf infants; the High Risk list and the later screening tests identified eight of them. Dr. Feinmesser concludes that in his situation the most effective screening will be done at the well-baby clinics.

Unfortunately, we in America do not have access to over 80 percent of the infant population at well-baby clinics, as Israel does. We must therefore look to the most effective way of identifying our deaf infants from information in the newborn nursery. The Jerusalem statistics do not give us too pessimistic an outlook for screening the High Risk Register infants: 90 percent of them were on the Register. We can well be guided by these results.

One other report should be mentioned: that of Dr. Erik Wedenberg of Stockholm, Sweden, found in Appendix B. Dr. Wedenberg is recognized as the father of modern infant screening, having reported as early as 1956 on neonatal screening. His paper is particularly pertinent to us in two ways: First, he gives us two types of behavioral responses in which he places confidence: the auro-palpebral reflex at 105 to 110 dB, and the arousal from sleep at levels as low as 70 to 75 dB. Second, the report on 11 children

Figure 1

UNIVERSITY OF COLORADO MEDICAL CENTER



Audiograms of B.J.,birth date 2/25/65. From birth to 2½years, observations of responses gave normal hearing range.

who had normal hearing at birth but whose hearing deteriorated rapidly at a later time. These were considered to be recessive deafnesses, a fact which again points to the possibility that the very hearing losses we are most interested in finding by mass screening may not be identifiable at birth.

In the outline which follows I have attempted to set up some guidelines which seem to follow from all that has been said here. The essence of it is that eternal vigilance should be observed in watching babies for deafness.

## Proposed Guidelines for Newborn Screening

I. Routine screening tests at birth using some form of behavioral response observations (or objective tests where they have been developed) on every infant on a High Risk Register for deafness, and follow-up monitoring of these infants.

A. Application of the High Risk Register in the following manner:[7]
  1. Through the managing physician, an examination of the medical records of all mothers and infants to determine pertinent historical facts.
  2. Physician-directed observation of all infants for physical findings and symptomatology which might be related to deafness.
  3. A questionnaire to be filled out by each mother, or an oral query, covering historical facts relating to deafness which might have been missed in the routine history.

B. Behavioral testing to be ideally accomplished in the following manner:
  1. Observations in a quiet room of the infants' responses in a manner specified by the professionals in charge, or to be later detailed at this conference.

II. Selected investigations on newborns who can be monitored developmentally and audiologically for at least a year. Proposed types of investigations to include:

A. Study following Dr. Derbyshire's design proposal.

B. Study of objective tests and their results as they relate to the child's later developmental status. These include:
  1. Cardiac Evoked Response Audiometry
  2. Crib-o-gram testing
  3. Evoked Response Audiometry
  4. Acoustic reflex testing

C. Study of Response Decrement testing to determine its effectiveness in identifying or predicting cerebral dysfunction.

III. Recommendations for physicians and all health personnel to attempt to identify the older deaf infant:

A. Some form of hearing test should be administered at every health visit.

B. At every health visit, the parent should be queried orally or by questionnaire as to the hearing status of the infant. (See Appendix C.)

C. When parents report doubts as to the hearing status of the child, every effort should be made to have an adequate hearing test performed.

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# EVALUATION OF METHODS FOR DETECTING HEARING IMPAIRMENT IN INFANCY AND EARLY CHILDHOOD

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L. Bauberger-Tell, Audiologist

Our screening program of the newborns extended over a period of 37 months, beginning on September 1st, 1967, and ending on September 30th, 1970.

The total number of screened newborns was 17,708. The number of "Apriton" positives (failures) was 309 (1.7 percent), 43 of these were also registered as "high risk." It should be pointed out that during the first six months (September 1967-February 1968) the percentage of "Apriton" positives (failures) was high – 7.5 percent. Since March 1968, after Mrs. Downs' visit, it gradually came down to 1 percent approximately. This decrease could be explained by the confidence gained by the nurses, briefed and guided by Mrs. Downs, as well as by the experience gained by the testers.

The number of "high risk" register was 3,547, which amounts to 20 percent of the total population of newborns.

When the "Apriton" positives were analyzed in relation to the infants "at risk", it was found that in the categories of "prematurity" and "deafness in family", the Apriton positives were significantly more frequent than in other categories.

All "Apriton" positives are being followed up, but here only the results of a 12-month period, beginning March 1968, will be analyzed. During that year (1968-69) 5,629 newborns were tested, and out of these, 66 or 1.1 percent were found "Apriton" positive. (Table 1) One died, three did not appear for further examination and could not be located, 51 were cleared when tested later in baby clinic, or in audiology clinic, or, sometimes, at home. Three were found profoundly deaf, two suspected of hearing impairment, three with brain damage, but with normal hearing, and the last three – with delayed speech but with normal hearing.

During the same period, out of 5,563 negatives (or cleared) on "Apriton", four appeared later to be deaf. These children will be reported on later.

Out of the above mentioned 5,629 newborns, 1,107 or 19.4 percent were registered as "high risk", and out of these, six were later identified as deaf. One infant, who was not "at risk" was later found to be deaf. (Table 2)

As was already pointed out earlier, 80-85 percent of the original newborn population visit the public baby clinics (Mother and Child Center).

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Table 1

NEWBORN SCREENING (APRITON)
12 months (1968 – 1969)

No. = 5,629

| SCREENING RESULTS | DIAGNOSIS | | TOTAL |
|---|---|---|---|
| | Deaf | Not Deaf | |
| Positives | 3 | 63 | 66 |
| Negatives | 4 | 5,559 | 5,563 |
| Total | 7 | 5,622 | 5,629 |

Table 2

HIGH RISK REGISTER
12 months (1968 – 1969)

No. = 5,629

| SCREENING RESULTS | DIAGNOSIS | | TOTAL |
|---|---|---|---|
| | Deaf | Not Deaf | |
| Positives | 6 | 1,101 | 1,107 |
| Negatives | 1 | 4,521 | 4,522 |
| Total | 7 | 5,622 | 5,629 |

Out of 5,629 newborns who were mentioned earlier, 4,860 (or 85 percent) visited the baby clinics, and 175 (3.5 percent) were found to be positive (failed the test).

All the positive children from the nurseries as well as from the Baby Clinics were followed up. About 50 percent of them visited our Audiology Center, and were tested when 7-10 months old. Others were tested when older, either in the Audiology Center, the Baby Clinics, or at home. We are continuing with our endeavour to follow up and test all of the children who failed.

About 50 percent of the original population continued to visit the Baby Clinic when 18-22 months old, and were evaluated on communication and developmental scale. Four percent of them failed and are being followed up at the Audiology Center, where their hearing is tested.

The children who are included in this program visit the Baby Clinic again when at the age of three years. We still don't know the numbers and the percentages of positives, but all who fail the hearing test will be retested at the Audiology Center.

As we found that a large percentage of children at this age are frequenting the kindergarten, we plan to visit them there. Thus we hope to reach most of the children who are included in this program and be able to determine the validity of the evaluation procedures performed in the previous stages.

Up to date, only nine children were identified as profound deaf. (Table 3) Five of them were positives (failed) on the "Apriton" test, while the other four were negatives (cleared) on "Apriton". Eight of these were registered as "high risk" – seven because of deafness in family and one because of consanguinity of parents.

In one male child who was negative on "Apriton", the deafness was familial, his two sisters being deaf. This child died at the age of one year. These children (including a distant cousin) suffered from renal tubular acidosis, and actually belong to a new but not yet described genetic syndrome of recessive deafness, associated with renal tubular acidosis.

Another child who was negative on "Apriton" received ototoxic drug, mainly ethacrynic acid, because of diabetes insipidus. This could explain his clearance at the nursery.

Our program of following up the children has not yet been completed. The youngest children are by now four months old and we believe that the final conclusions could be drawn only when they reach the age of three to four years.

Nevertheless, some comments can be given and conclusions arrived at:

Early detection of hearing impairment is very important for the success of habilitation of the deaf child. It is of general opinion that the identification of deafness during the first months of life will permit starting with educational treatment, which will

Table 3

DEAF CHILDREN (1 YEAR OR MORE)

| CHILD | APRITON | HIGH RISK | REMARKS |
|---|---|---|---|
| 1. O.S. | + | Deafness,<br>Familial | |
| 2. Sh.G. | – | Consanguinity<br>parents | Diabetes (insip.) fam.<br>Ototoxic drug<br>Perceptual disturbance |
| 3. H.Sh. | – | Deafness, fam.<br>Consang., parents | |
| 4. L.M. | – | Deafness,<br>Familial | |
| 5. S.I. | + | Deafness,<br>familial | |
| 6. S.Sh. | + | Deafness,<br>Familial | Deafness with albinism<br>Sex-linked syndrome |
| 7. L.A. | – | Deafness,<br>Familial | Renal tubular<br>acidosis, famil. +<br>deafness. Exitus |
| 8. C.E. | + | – | Mental retardation |
| 9. H.E. | + | Deafness,<br>Familial | |

provide the deaf child with the optimum conditions for the acquisition of language and normal development. The newborn nursery is the only place where the child population as a whole is available for screening and it would be ideal if the child who is born deaf could be identified there.

Auditory screening programs have been implemented in newborn nurseries in different countries and in two such programs in Israel, one in Haifa and the other in Jerusalem. The Jerusalem program is based on mass hearing testing of the newborn, and selective screening of infants registered as "high risk" for deafness.

In addition, both in Haifa and in Jerusalem, a method of screening of all children attending the Baby Clinics (Mother and Child Welfare Clinics) was investigated.

Validity of screening any disease in general, and deafness in particular, depends on the sensitivity of the diagnostic procedure.

Sensitivity means the ability of the procedure to give a positive finding when the person evaluated has the disease under study. A sensitive hearing test will detect nearly all cases of deafness with a low rate of positive errors. A sensitive high risk register should also detect for further identification most of the hearing impaired children with small proportion of errors.

Our program, as was pointed out, is not yet accomplished, the children are still being followed up, but from the results already obtained it seems that the "Apriton" screening test is not very sensitive and, therefore, the validity of the test is questionable. The 1 percent of positives is acceptable for screening purposes, but the negative errors or false negatives is relatively high.

Out of the nine children who were identified as deaf, four were Apriton negative at birth, and considering that one of these four was not congenitally deaf, yet the test is not sensitive enough.

The "high risk" register is of limited value. In spite of the enthusiasm at the beginning, several years ago, with the "high risk" register, the results have been disappointing, and there is now wide acceptance of the need for careful reappraisal. If too many are put on the register, the arrangements for follow-up become unmanageable; if too few, or the wrong babies are selected, and handicapped children are found not to have been registered, the scheme fails.

Rogers seriously doubts the value of the "risk register" for all kinds of handicaps. Others criticize the ill-defined and imprecise risk categories, tendency to omit social factors and to neglect children not on the register.

Though in our program, which is still continued, eight out of nine at-risk children were identified as deaf, in Haifa, among 10,000 children screened, 13 profound congenital deaf were found, nine of whom had been considered "risk" cases and four not to be "at risk". Even though deafness among "at risk" groups is more common than in the ordinary population, on the other hand – the "high risk" register has too many false positives, which means that the number of registered "at risk" and later proved to be of normal hearing is too high to be selective, and the number of false negatives is also relatively high (in the Haifa program).

Another point which should be considered is the practicability of the evaluation procedure when expenditure and economics play an important role. In our program, the expenses were high. No volunteers were engaged. A full-time audiologist was necessary for briefing and supervising the work of the tester, as well as following up the positive cases. Last but not least, the expenses for the statistical work-up are not negligible.

However, we completely agree with Mrs. Downs that such a program offers valuable information in the etiology of hearing impairment and induces awareness in doctors, nurses and educators of hearing problems. Valuable information is also acquired on early language behavior of the deaf child.

Since in our program seven out of nine deaf children have a history of deafness in the family we decided to continue to screen all the newborns at the Hadassah Hospital who have a family history of deafness, or present evident symptoms of multiple handicaps. This procedure is economical and we hope that with the completion of our program and the collection of data, another category of significance will be introduced.

Another place where a large part of the infant population can be screened was the Baby Clinic. In Israel, about 85 percent of the child population visits the Baby Clinics during the first months of life, and those visits are often repeated monthly. The child's hearing is tested when 5-6 months old; the testing procedure lasts approximately five minutes, and forms an integral part of the general growth and development evaluation program. It is very economical, as no additional staff is required for the hearing assessment. The child's response is easy to observe, as it consists of the turn of the head to the source of sound.

About 2 percent of the children (the percentage in the later period) who fail the test are later tested in the Audiology Center in free field condition and those suspected of hearing impairment are followed up.

The test is relatively sensitive and has the advantage of detecting hearing impairment of those with less than profound deafness. Thus in some children who were identified as hearing impaired, otitis media was found, and when treated – their hearing impairment disappeared.

The process of auditory maturation is unknown but it is feasible that at the age of six months the auditory pathways achieve relative maturity. At this age, the reaction to the auditory stimulus is no more a reflex, and cases with central damage may be detected.

We believe that the screening in the Baby Clinic is effective and practicable in Israel. In other countries where many children do not visit public clinics, general practitioners and pediatricians should include the testing of hearing as a routine evaluation of the development of the child.

## Final Comments

1. The conclusions as to the validity of the screening methods as applied by us will be drawn only when the program has been completed and the children followed up until 3-4 years of age.

2. At the present time, it seems that the Apriton test is not sensitive enough and the screening value of the newborns by this procedure is questionable.

3. The value of the "at risk" register in its present form is doubtful.

4. Effort should be made to find a more sensitive method for screening newborns for the detection of hearing impairment.

5. The screening of children for hearing impairment at the Baby Clinics seems to be of great value, practical and economical. If no screening opportunities arise, the hearing and communication ability should be included in the routine evaluation of the development of the child.

# AUDITORY TESTS ON NEWBORN INFANTS

Erik Wedenberg, M.D.*

For measuring the hearing of newborn infants it is necessary to use objective methods. I have performed two kinds of objective hearing tests (Wedenberg, 1956, 1963).

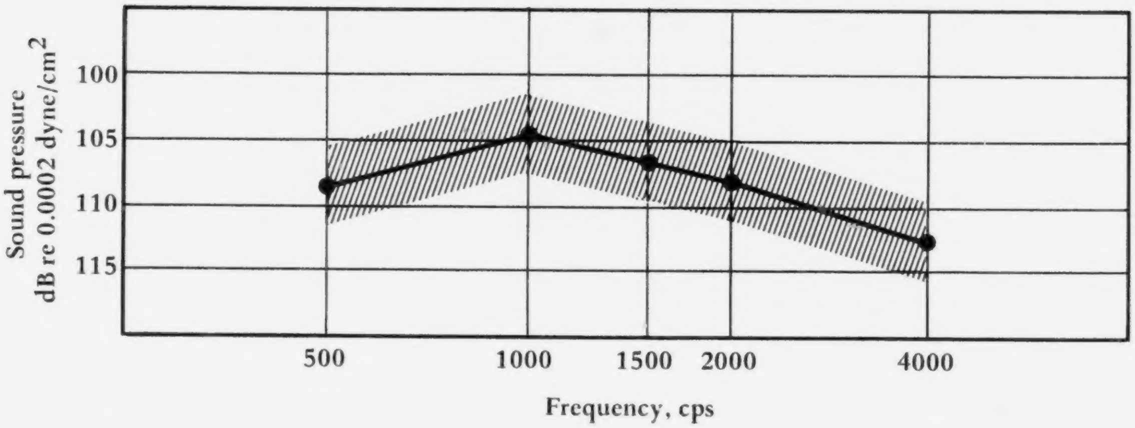
I. Threshold determination of the auropalpebral reflex (APR) showing the contraction of the orbicularis oculi muscle.

II. Determination of the intensity of sounds required to waken the child who is in a certain depth of sleep. A specially designed tone audiometer was used for these tests.

Those tests are conducted with the child lying on its side in a cot. In the first 20 subjects, who had been selected as probably being normal, the APR was elicited at a threshold of 105-115 dB for all the frequencies tested in the range 500-4,000 cps. (Figure 1)

Figure 1

MEAN THRESHOLD CURVE FOR THE AURO-PALPEBRAL REFLEX OF NEWBORN INFANTS



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That it is possible to use these threshold determinations of APR as a test of hearing level in the *newborn* is due to the similarity between APR and another acoustic reflex, the stapedius reflex; this similarity is familiar in both normals and persons with defective hearing. The APR reflex-threshold curves for *adults with normal hearing* are very similar; also from the anatomical aspect the two reflexes have much in common. The afferent part (acoustic nerve) and the efferent part (facial nerve) of the reflexes are common to both, but the center of the stapedius reflex is situated in the pons and that of the APR in the reticular formation.

The APR threshold curve for the children examined should indicate normal hearing. In the individual case, however, the fact that the APR has been elicited at a particular frequency with a tone of "normal" intensity cannot be taken as *proof* that the hearing for the frequency in question is normal. As has been shown in the case of the stapedius reflex, there may be severe hearing loss with recruitment. So in order to distinguish between normal hearing and impaired hearing with recruitment, experiments were performed to find the intensity required to awaken a child in a depth of sleep such that the APR could be elicited by tactile stimulus. The audiometer was the same as that used in the APR tests. The frequencies were 500 and 3,000 cps, and the tones were of 1-5 seconds duration, emitted irregularly for one minute. At an intensity of 70-75 ds (dB), if not before, all the infants showed signs of waking such as a change in the breathing rhythm or the flickering of the eyelids.

As an aid in distinguishing between normal hearing and different types of hearing impairment, a chart was compiled showing how a normal child and one with impaired hearing might be supposed to react to APR and awaking from sleep. (Figure 2)

The case on the following page shows my mode of procedure. (Figure 3)

Case 8 J.T. b. 9.10.57

**Heredity.** – Third child of Rh immunized mother.

**Peri-natal status.** – Jaundice on third day, opisthotonus posture. Bilirubin 22.3 mg per 100 ml. No Moro embrace or grasp reflexes. The reflexes gradually recovered.

**General diagnosis.** – Neonatal hemolytic disease, nuclear jaundice; hearing impairment?

**Original hearing test.** – At 14 days: bilateral retrocochlear loss.

**Follow-up test.** – At 2½ and 3 4/12 years: bilateral retrocochlear loss.

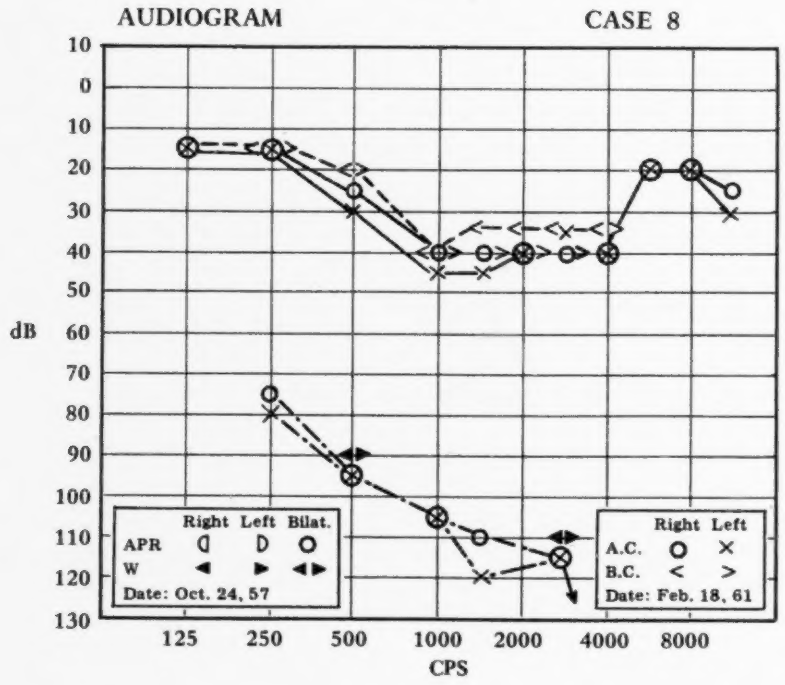
**Comments.** – According to the chart in Figure 2 the absence of APR but waking at higher intensities than 70-75 dB at 500 and 3,000 cps indicates either conductive or retrocochlear loss. The former is ruled out by the ear examination which disclosed normal conditions; the diagnosis is therefore retrocochlear hearing loss, and this is compatible with the localization of the hearing defect in neonatal hemolytic disease,

Figure 2

PROSPECTIVE SCHEDULE OF DIFFERENT KINDS OF HEARING LOSS

| | Normal Hearing | Cochlear Hearing Loss | Conductive Hearing Loss<br>Retrocochlear Hearing Loss | Total Deafness<br>Severely Hard of Hearing |
|---|---|---|---|---|
| Auropalpebral Reflex | + | + | – | – |
| Waking ≤70-75 dB | + | – | – | – |
| Waking >70-75 dB | + | + | + | – |

Figure 3



A.C.: Air Conduction
B.C.: Bone Conduction
W.: Waking

**Dash-dotted curve: -·—·— Stapedius reflex bilateral Feb. 16, 61**

namely the dorsal and ventral nucleus in the medulla oblongata. Since, in retrocochlear impairment, the loudness increases parallel with the loudness perception of the person with normal hearing, waking at 500 cps and 95 dB indicates a hearing loss of 95 less 70, or 25 dB, and waking at 3,000 cps and 105 dB a loss of 105 less 70, or 35 dB. At the examination at 2½ years this child could perceive the s-sound well at several meters; this implies fairly good hearing above 4,000 cps, beyond which frequency the main formants of the s-sound lie. *In this case an audiogram could be recorded at the first test performed when the child was 14 days old.*

In Sweden this method is in widespread use, but only for neonatal testing of children belonging to the high risk group. I think testing all children at birth is too time consuming considering the rarity of deafness at birth (0.7 percent).

These objective hearing tests are extremely reliable. But sometimes it happens that children, whose hearing at birth is determined as normal and at control measurements at different ages is found to be normal, suddenly get a high frequency hearing loss without any known cause. These hearing impairments certainly have an endogenous origin. In many cases it has been possible to detect clinical or subclinical hearing defects in one or both parents who therefore could be identified as gene carriers (Anderson & Wedenberg, 1968, 1970). The peculiarities of the hearing of the parents are small but distinct dips in the middle frequency range of the hearing threshold and abnormally high thresholds for the acoustically elicited stapedius reflex.

The progressivity of the hearing loss found in their children has been, of the families I studied, all eleven children had normal hearing at birth but at about one year of age the deterioration began and now they are all severely hard of hearing (Barr & Wedenberg, 1965).

As to a patient with an endogenous hearing defect an audiogram from one occasion is like a single frame from a whole film sequence, which stubbornly continues its course at a gene-controlled predestined speed.

A reverse condition has also been reported by some authors. Neonatal hearing tests indicate deafness but hearing tests at later occasions show normal hearing. These findings of so-called false positive tests are probably due to technical errors. I only know one type of case who with certainty is deaf at birth, but who later on has normal hearing. All such children have severe neonatal jaundice. In the Karolinska Hospital we have followed many such cases. Before the blood exchange, which is made when the bilirubin is > 20 mg per 100 ml, the APR cannot be elicited and waking with sound stimulation is impossible. After the blood exchange the children gradually begin to react to auditory stimulation and the APR and waking tests become normalized. When the children have grown up their hearing tests show normal values. The explanation of this remarkable change is the following:

We know that a causal connection exists between neonatal jaundice (kernicterus) and hearing impairment. The buildup of bilirubin in the cochlear nuclei brings about a disturbance in the metabolism of these cells usually with cell destruction and accompanying deafness. Thanks to an early blood exchange it is now possible to prevent the cell destruction in the cochlear nuclei and escape hearing impairments. In other words, the condition is reversible.

In the work with intrauterine measurements of tone response in the human fetus (Johanson, Wedenberg, Westin), which work has now continued during six years, we have found fetuses who have not responded to the presented tones. I have tested their hearing after birth with the result: total deaf or severely hard of hearing. But in one case we have shown that a child before birth had responded normally to the tones applied, and after birth in several measurements had been determined as normal hearing but suddenly at five years of age got a hearing defect. In this case the parents have been identified as gene carriers. In the Department of Audiology, Karolinska Hospital, Stockholm, we now work with the problem of distinguishing already at birth such children, who seem to respond normally to our neonatal hearing tests but nevertheless after some time get a hearing loss of endogenous origin. This year I hope we shall publish our findings.

The endogenous hearing defects that appear in earlier infancy of human beings have their parallels among animals. It has been known for a long time that certain races of animals (rats, guinea pigs) can hear when they are born, but lose their hearing capacity during their first month of life. Histological studies have shown that in these animals the cochlea is normally developed at birth, but degenerates gradually (Grüneberg et al., 1940).

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## PARENT QUESTIONNAIRE

*(Answer Yes if you have very definitely seen your child do these things even once or twice. If you have never seen him do them, answer No.)*

| | YES | NO |
|---|---|---|
| **BIRTH TO FOUR MONTHS** | | |
| 1. Does he startle to a sudden sound such as a cough, a shout, a dog bark, or a handclap? (Discount responses to a door slamming, the stamp of a foot, a loud airplane or truck noise, and other vibrations.) | ___ | ___ |
| 2. When he is sleeping in a quiet room, does he stir or awaken, when someone speaks or a noise is made near him? (Some babies are constantly in noisy surroundings, and such infants tend to inhibit their responses unless they have been in quiet for some time.) | ___ | ___ |
| 3. When he is crying or fretful, does he appear to calm down even momentarily when you speak out of eye-shot or when music starts up or when a sudden loud noise occurs? | ___ | ___ |
| 4. At three to four months, does he occasionally seem to make feeble beginning head-turn toward a sound, or move his eyes in its direction? | ___ | ___ |
| **4 - 8 MONTHS** | | |
| 1. Does he turn his head and eyes toward a sound on one side of him that is out of his peripheral vision? (At 4 months, he should begin to turn directly to the side.) | ___ | ___ |
| 2. In a quiet situation, does he change expression or widen his eyes when he hears a fairly loud sound or voice? | ___ | ___ |
| 3. Does he briefly enjoy ringing bells or squeezing noise-makers or shaking a rattle? | ___ | ___ |
| 4. By six months, does he seem to talk or babble to persons in response to their speaking or making noises? | ___ | ___ |
| 5. By six months, does his babbling include four different sounds? (Although a deaf baby's babbling sounds just like a normal hearing baby the first few months, by six months he usually uses only one or two gross vowel sounds.) | ___ | ___ |
| **8 - 12 MONTHS** | | |
| 1. Does he turn directly and quickly toward an interesting soft noisemaker or to his name called or to a "sh-sh" out of his peripheral vision? | ___ | ___ |
| 2. Does he use different pitches in his babbling? | ___ | ___ |
| 3. Does he make several different consonant sounds in his babbling? | ___ | ___ |
| 4. Does he seem to enjoy music and respond to it by listening or bouncing or vocalizing? | ___ | ___ |

*If you have answered No to any of these questions at the present age level of your child, his hearing should be checked at an Audiology Clinic.*

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* Univ. of Colo. Med. Center, Div. of Otolaryngology, M. Downs.

## QUESTIONNAIRE*

## NEONATAL AUDITORY SCREENING

There is considerable interest on the part of audiologists, otologists, pediatricians and public health officials in the subject of neonatal auditory screening. In spite of much work by many individuals some aspects of this program remain controversial. Recognizing that these issues must ultimately be resolved on the basis of adequate factual data accumulated by controlled studies, it is nevertheless of some interest to obtain a cross section of opinion based on personal knowledge, experience, reasoning and informally exchanged information from individuals, such as yourself, with some interest and competence in the general area of hearing evaluation and conservation in children. As background information for a conference to be held later this year, we would like to impose on your time to ask you to check your response to the following questions, and return it to us as soon as possible. If you have no experience or particular interest in this area of newborn screening, simply indicate that fact and we will remove your name from our mailing list.

1. Are you in favor of newborn screening by any currently reported method for high risk newborns prior to discharge? ☐ Yes ☐ No
2. Are you in favor of newborn screening by any currently reported method for all newborns prior to discharge? ☐ Yes ☐ No
3. Do you feel that hearing defects would best be detected by screening
   - ☐ in the newborn period
   - ☐ in the first six months
   - ☐ before one year of age
   - ☐ before two years of age
   - ☐ before school entrance
4. Which of the currently available screening methods do you feel is most promising?

   ______________________________
5. False positives, i.e., infants with normal hearing who fail to pass screening test, would render a testing method unsatisfactory if they exceeded _____ per 10,000 newborns tested.
6. If you were referred an infant under three months of age with suspected hearing loss, could you obtain appropriate diagnostic services to confirm or rule out this diagnosis? ☐ Yes ☐ No
7. If you were referred an infant under three months of age with suspected hearing loss, could you obtain appropriate rehabilitative or treatment service? ☐ Yes ☐ No
8. Early medical treatment is effective in preventing hearing loss or improving functional hearing in approximately ______% of the infants failing screening.
9. Early auditory habilitation and special education is effective in ______ %.
10. At what age would you first consider the use of a hearing aid?

**PLEASE RETURN TO:**
George C. Cunningham, M.D., Chief
Bureau of Maternal and Child Health
California State Department of Public Health
2151 Berkeley Way
Berkeley, California 94704

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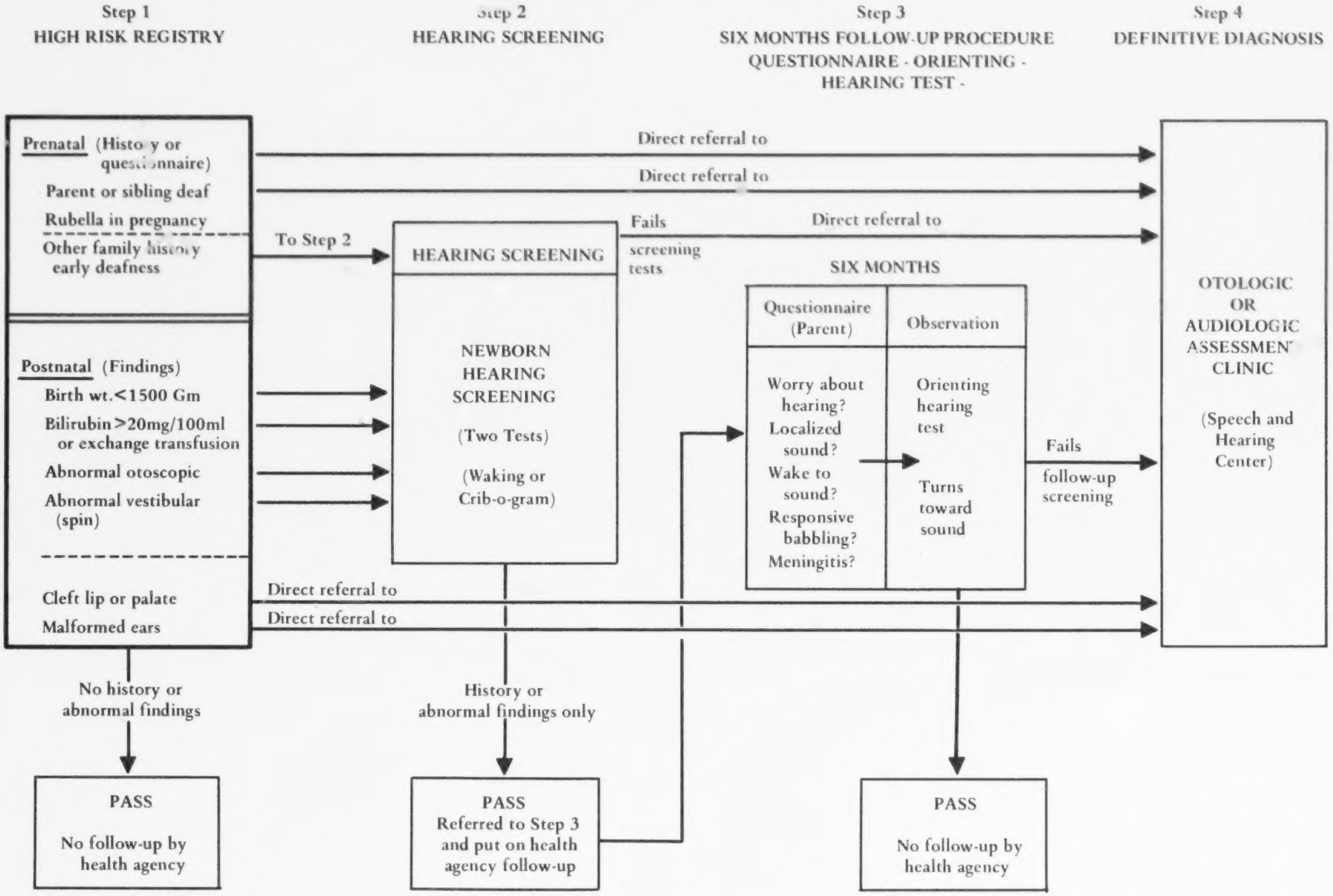
* Sent to pediatricians and otolaryngologists listed in the Directory of Medical Specialists, and to audiologists, in the following cities in California: Redding, San Francisco, Fresno, Los Angeles, San Diego, in June 1970.

**APPENDIX II**

## TALLY OF RESPONSES TO NEONATAL AUDITORY SCREENING QUESTIONNAIRE

Mailed Out June 1970

| AUDIOLOGISTS | PEDIATRICIAN | OTOLOGISTS | | | |
|---|---|---|---|---|---|
| 19 | 69 | 23 | Yes | | (1) Are you in favor of newborn screening by any currently reported method for all HIGH RISK newborns prior to discharge? |
| 2 | 3 | 3 | No | | |
| - | 6 | 2 | No Response | | |
| 12 | 40 | 14 | Yes | | (2) Are you in favor of newborn screening by any currently reported method for ALL newborns prior to discharge? |
| 7 | 31 | 12 | No | | |
| 2 | 7 | 2 | No Response | | |
| 4 | 15 | 8 | In the Newborn Period | | (3) Do you feel that hearing defects would best be detected by screening? |
| 11 | 30 | 10 | In the First 6 Months | | |
| 2 | 9 | 2 | Before 12 Months | | |
| 1 | 5 | 4 | Before 24 Months | | |
| 3 | 6 | - | Before School Entrance | | |
| - | 1 | - | Qualified | | |
| - | 10 | 4 | No Response | | |
| 8 | 14 | 4 | Calibrated Sounds, i.e., Apriton, Warblett, etc. | | (4) Which of the currently available screening methods do you feel is most promising? |
| 4 | 5 | 2 | Observations | | |
| | 2 | 1 | Responses to Cross Stimuli, i.e., Clapper, Bells, etc. | | |
| | 3 | 6 | EEG Audiometry | | |
| | 3 | - | PGR (EDR) Audiometry | | |
| 1 | 2 | - | Evoked Potential, e.g., Heart Rate | | |
| | | 2 | Reflex, i.e., Moro | | |
| | 2 | 3 | Rely on Audiologist | | |
| | | - | None are Adequate | | |
| 1 | 2 | 3 | Qualified Comment | | |
| 7 | 46 | 8 | No Response | | |
| 1 | 20 | 7 | 2 - 50 | NUMBER FALSE POSITIVES PER 10,000 | (5) False positive, i.e., infants with normal hearing who fail to pass screening test, would render a testing method UNSATISFACTORY if they exceeded _____ per 10,000 newborns tested. |
| - | 6 | 2 | 100 | | |
| 1 | 4 | - | 200 | | |
| 1 | 3 | - | 300 | | |
| 10 | 8 | 7 | 500 - 1500 | | |
| - | - | - | 3000 | | |
| 1 | - | - | Qualified | | |
| 7 | 35 | 12 | No Response | | |
| 17 | 63 | 19 | Yes | | (6) If you were referred an infant under 3 months of age with suspected hearing loss, could you obtain appropriate diagnostic services to confirm or rule out this diagnosis? |
| 2 | 8 | 6 | No | | |
| 2 | 7 | 3 | No Response | | |
| 18 | 53 | 17 | Yes | | (7) If you were referred an infant under 3 months of age with suspected hearing loss, could you obtain appropriate rehabilitative or treatment service? |
| 2 | 13 | 8 | No | | |
| 1 | 12 | 3 | No Response | | |
| 3 | 10 | 9 | 0 - 40 | PERCENT TREATMENT EFFECTIVE | (8) Early medical treatment is effective in preventing hearing loss or improving functional hearing in approximately _____ % of the infants failing screening? |
| 2 | 10 | 2 | 50 - 70 | | |
| 1 | - | 2 | 80 - 90 | | |
| 1 | 1 | 2 | Over 90 | | |
| 1 | 2 | - | Qualified | | |
| 13 | 58 | 13 | No Response | | |
| 2 | 8 | 4 | 0 - 50 | PERCENT SPECIAL TRAINING EFFECTIVE | (9) Early auditory habilitation and special education is effective in _____%. |
| 1 | 2 | 3 | 50 - 70 | | |
| - | 13 | 5 | 70 - 90 | | |
| 6 | 7 | 6 | Over 90 | | |
| - | 3 | - | Qualified | | |
| 10 | 48 | 13 | No Response | | |
| 18 | 37 | 10 | Birth - 6 Months | AGE IN MONTHS | (10) At what age would you first consider the use of a hearing aid? |
| 2 | 16 | 2 | 7 - 18 Months | | |
| - | 4 | 12 | 24 - 36 Months | | |
| - | 2 | - | 42 Months or More | | |
| 1 | 3 | - | Qualified | | |
| - | 15 | 4 | No Response | | |



APPENDIX III. SUGGESTED FLOW CHART FOR NEWBORN HEARING SCREENING PROGRAM WITH FOLLOW-UP.
*(Adapted from schema developed by Isabelle Rapin, M.D., 1/25/71).*